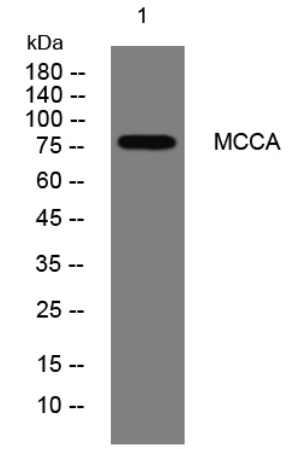
MCCA rabbit pAb

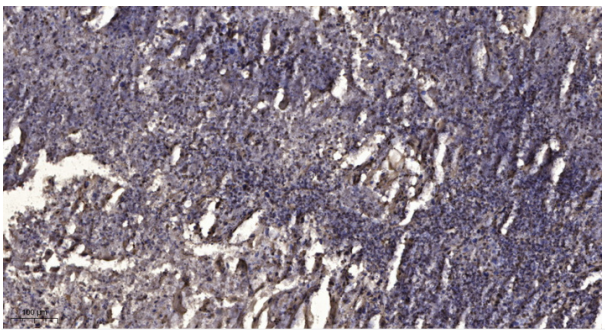
Catalog No	BYab-11767
Isotype	IgG
Reactivity	Human; Mouse;Rat
Applications	WB;IHC
Gene Name	MCCC1 MCCA
Protein Name	MCCA
Immunogen	Synthesized peptide derived from human MCCA AA range: 620-670
Specificity	This antibody detects endogenous levels of MCCA at Human/Mouse/Rat
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit serum by affinity-chromatography using specific immunogen.
Dilution	WB 1:500-2000;IHC-p 1:50-300
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

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.

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).