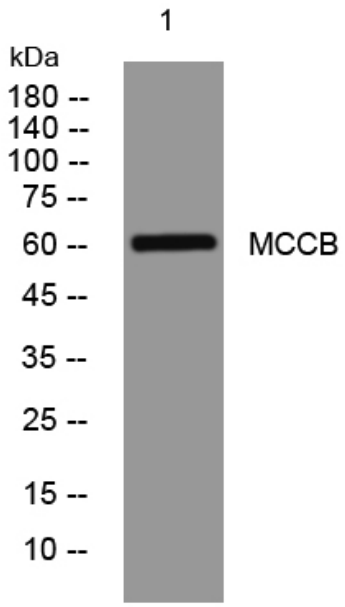
MCCB rabbit pAb

Catalog No	BYab-07994
Isotype	IgG
Reactivity	Human; Mouse;Rat
Applications	WB
Gene Name	MCCC2 MCCB
Protein Name	MCCB
Immunogen	Synthesized peptide derived from human MCCB AA range: 204-254
Specificity	This antibody detects endogenous levels of MCCB at Human/Mouse/Rat
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.108% 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
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	Methylcrotonoyl-CoA carboxylase beta chain, mitochondrial (MCCase subunit beta) (EC 6.4.1.4) (3-methylcrotonyl-CoA carboxylase 2) (3-methylcrotonyl-CoA carboxylase non-biotin-containing subunit) (3-methylcrotonyl-CoA:carbon dioxide ligase subunit beta)
Observed Band	60kD
Cell Pathway	Mitochondrion matrix .
Tissue Specificity	Testis,Uterus,
Function	catalytic activity:ATP + 3-methylcrotonoyl-CoA + HCO(3)(-) = ADP + phosphate + 3-methylglutaconyl-CoA.,disease:Defects in MCCC2 are the cause of methylcrotonoyl-CoA carboxylase deficiency type 2 (MCC2 deficiency) [MIM:210210]. MCC2 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:Belongs to the accD/PCCB family.,similarity:Contains 1 carboxyltransferase domain.,subunit:Probably a dodecamer composed of six biotin-containing alpha subunits and six beta subunits.,
Background	This gene encodes the small 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 3T3 cells, primary antibody was diluted at 1:1000, 4°over night