Best Tools for immunology Research

MCCA rabbit pAb

Catalog No :	YT7132
Reactivity :	Human;Mouse;Rat
Applications :	WB;IHC
Target :	MCCA
Fields :	>>Valine, leucine and isoleucine degradation;>>Metabolic pathways
Gene Name :	MCCC1 MCCA
Protein Name :	MCCA
Human Gene Id :	56922
Human Swiss Prot No :	Q96RQ3
Mouse Gene Id :	72039
Mouse Swiss Prot No :	Q99MR8
Rat Gene Id :	294972
Rat Swiss Prot No :	Q5I0C3
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
Dilution :	WB 1:500-2000;IHC 1:50-300
Purification :	The antibody was affinity-purified from rabbit antiserum by affinity-

chromatography using epitope-specific immunogen.

Concentration : 1 mg/ml

Storage Stability : -15°C to -25°C/1 year(Do not lower than -25°C)

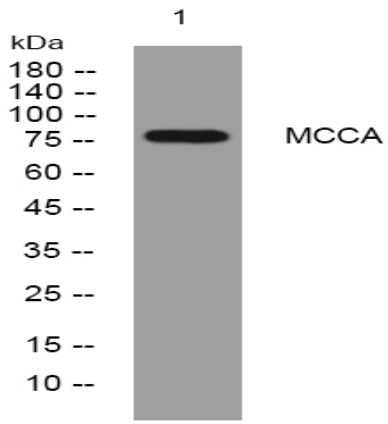
Molecularweight : 80kD

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],

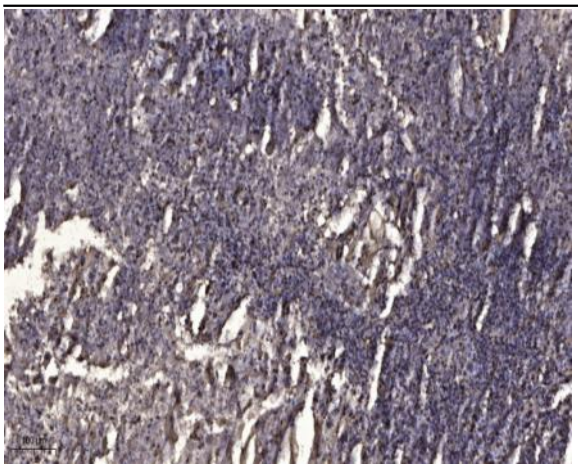
Function : catalytic activity:ATP + 3-methylcrotonoyl-CoA + HCO(3)(-) = ADP + phosphate + 3-methylglutaconyl-CoA.,cofactor:Biotin.,disease:Defects in MCCC1 are the cause of methylcrotonoyl-CoA carboxylase deficiency type 1 (MCC1 deficiency) [MIM:210200]. MCC1 deficiency is an autosomal recessive disorder 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

Subcellular Location : Mitochondrion matrix .

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).