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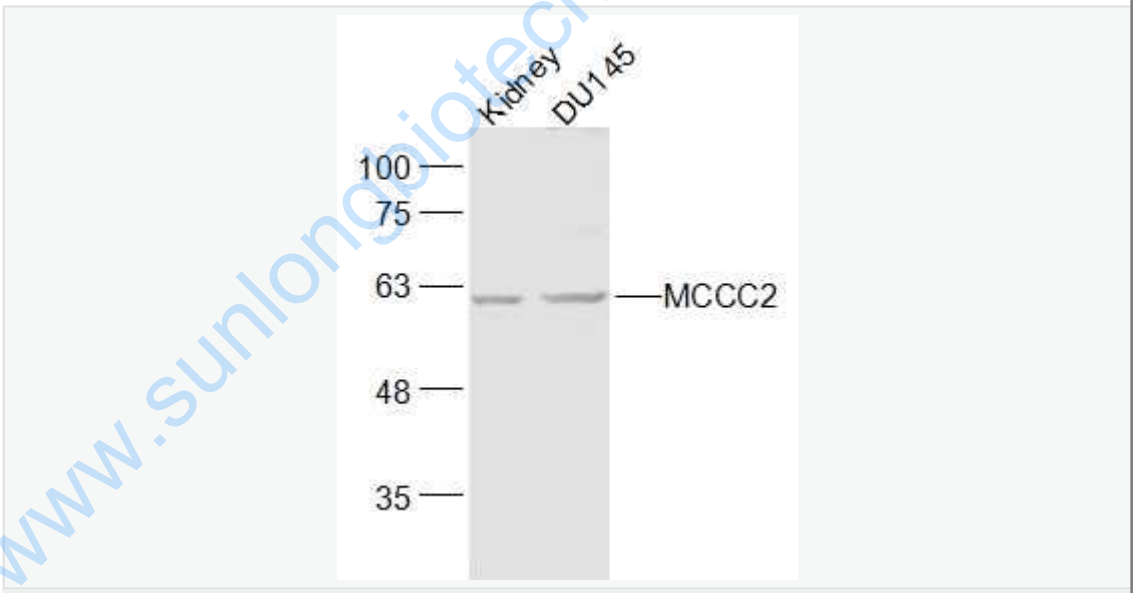
Rabbit Anti-MCCC2 antibody

SL18719R

Product Name:	MCCC2
Chinese Name:	MCCC2蛋白抗体
Alias:	3 methylcrotonyl CoA carboxylase 2; 3 methylcrotonyl CoA carboxylase non biotin containing subunit; 3 methylcrotonyl CoA:carbon dioxide ligase subunit beta; 3-methylcrotonyl-CoA carboxylase 2; 3-methylcrotonyl-CoA carboxylase non-biotin-containing subunit; 3-methylcrotonyl-CoA:carbon dioxide ligase subunit beta; Biotin carboxylase; MCCase subunit beta; MCCB; MCCB_HUMAN; MCCC 2; Mccc2; Methylcrotonoyl CoA carboxylase 2 (beta); Methylcrotonoyl CoA carboxylase beta chain mitochondrial; Methylcrotonoyl Coenzyme A carboxylase 2 (beta); Methylcrotonoyl-CoA carboxylase beta chain; mitochondrial; Non biotin containing subunit of 3 methylcrotonyl CoA carboxylase.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,Xenopus tropicalis
Applications:	WB=1:500-2000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	61kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human MCCC2:351-450/563
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed

Product Detail:	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] **Function:** Carboxyltransferase subunit of the 3-methylcrotonyl-CoA carboxylase, an enzyme that catalyzes the conversion of 3-methylcrotonyl-CoA to 3-methylglutaconyl-CoA, a critical step for leucine and isovaleric acid catabolism. **Subunit:** Probably a dodecamer composed of six biotin-containing alpha subunits (MCCC1) and six beta (MCCC2) subunits. **Subcellular Location:** Mitochondrion matrix **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. **Similarity:** Belongs to the AccD/PCCB family. Contains 1 carboxyltransferase domain. **SWISS:** Q9HCC0 **Gene ID:** 64087 **Database links:** Entrez Gene: 64087 Human Entrez Gene: 78038 Mouse Entrez Gene: 361884 Rat Omim: 609014 Human

	SwissProt: Q9HCC0 Human SwissProt: Q3ULD5 Mouse SwissProt: Q5XIT9 Rat Unigene: 604789 Human Unigene: 137327 Mouse Unigene: 33635 Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Sample: Kidney (Mouse) Lysate at 40 ug DU145(Human) Cell Lysate at 30 ug Primary: Anti-MCCC2 (SL18719R) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 61 kD

	Observed band size: 61 kD