

Human Methylcrotonoyl-CoA Carboxylase beta Chain, Mitochondrial (MCCC2) ELISA Kit | MCCC2 elisa kit

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CATALOG # AAA331846	REACTIVITY Human
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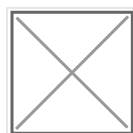
PRODUCT SYNONYMS

Methylcrotonoyl-CoA Carboxylase beta Chain; Mitochondrial (MCCC2); Human Methylcrotonoyl-CoA Carboxylase beta Chain; Mitochondrial (MCCC2) ELISA Kit; MCCC2; MCCB; biotin carboxylase; non-biotin containing subunit of 3-methylcrotonyl-CoA carboxylase; MCCC2 elisa kit

PRODUCT SPECIFICATIONS

Reactivity	Human
Specificity	Human Methylcrotonoyl-CoA carboxylase beta chain, mitochondrial (MCCC2) ELISA Kit has high sensitivity and excellent specificity for detection of Human MCCC2. No significant cross-reactivity or interference between Human MCCC2 and analogues was observed.
Sequence Length	563
Assay Type	Sandwich ELISA (Quantitative)
Samples	Cell Culture Supernatants, Serum, Plasma, Other Biological Fluids
Detection Method	Colorimetric
Usage Notes	* Do not mix components from different kit lots or use reagents beyond the kit expiration date. * Allow all reagents to warm to room temperature for at least 30 minutes before opening. * Pre-rinse the pipet tip with reagent, use fresh pipet tips for each sample, standard and reagent to avoid contamination. * Unused wells must be kept desiccated at 4 degree C in the sealed bag provided. * Mix Thoroughly is very important for the result. It is recommended using low frequency oscillator or slight hand shaking every 10 minutes. * It is recommended that all samples and standards be assayed in duplicate or triplicate.
Preparation and Storage	Store at 2-8 degree C.

IMAGES



Standard Curve (Sample)

Fig.1. Human Methylcrotonoyl-CoA carboxylase beta chain, mitochondrial (MCCC2) Standard Curve.

ADDITIONAL INFORMATION

Related Product Information for MCCC2 elisa kit	Description: This Human Methylcrotonoyl-CoA carboxylase beta chain, mitochondrial (MCCC2) ELISA Kit employs a two-site sandwich ELISA to quantitate MCCC2 in samples. An antibody specific for MCCC2 has been pre-coated onto a microplate. Standards and samples are pipetted into the wells and any MCCC2 present is bound by the immobilized antibody. After removing any unbound substances, a biotin-conjugated antibody specific for MCCC2 is added to the wells. After washing, Streptavidin conjugated Horseradish Peroxidase (HRP) is added to the wells. Following a wash to remove any unbound avidin-enzyme reagent, a substrate solution is added to the wells and color develops in proportion to the amount of MCCC2 bound in the initial step. The color development is stopped and the intensity of the color is measured. Background: MCCC2 is 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. Defects in MCCC2 are the cause of methylcrotonoyl-CoA carboxylase deficiency type 2 (MCC2 deficiency). 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.
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NCBI AND UNIPROT INFORMATION

NCBI GI #	11545863
NCBI GeneID	64087
NCBI Accession #	NP_071415.1
NCBI GenBank Nucleotide #	NP_071415.1
UniProt Accession #	Q9HCC0
NCBI Official Full Name	methylcrotonoyl-CoA carboxylase beta chain, mitochondrial isoform 1
NCBI Official Synonym Full Names	methylcrotonoyl-CoA carboxylase 2
NCBI Official Symbol	MCCC2
NCBI Official Synonym Symbols	MCCB
NCBI Protein Information	methylcrotonoyl-CoA carboxylase beta chain, mitochondrial
UniProt Protein Name	Methylcrotonoyl-CoA carboxylase beta chain, mitochondrial
UniProt Gene Name	MCCC2
UniProt Synonym Gene Names	MCCB; MCCase subunit beta
UniProt Entry Name	MCCB_HUMAN

For Research Use Only. Not for use in diagnostic or therapeutic procedures. Small volumes of product may become entrapped in the seal of the vial during shipment and storage; if necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the cap.