

## Rabbit Anti-MCEE antibody

SL18720R

Product Name:	MCEE
Chinese Name:	MCEE蛋白抗体 MCEE蛋白抗体
Alias:	DL methylmalonyl CoA racemase; DL-methylmalonyl-CoA racemase; EC 5.1.99.1; GLOD2; Glyoxalase domain containing 2; MCEE; MCEE_HUMAN; Methylmalonyl CoA epimerase; Methylmalonyl-CoA epimerase; methylmalonyl-CoA epimerase, mitochondrial; mitochondrial; OTTHUMP00000160122.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human, Mouse, Rat, Chicken, Dog, Horse,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100- 500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	15kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
<b>Concentration:</b>	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human MCEE:101-176/176
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:	MCEE is a 176 amino acid mitochondrial protein that belongs to the glyoxalase I family. MCEE catalyzes the interconversion of D- and L-methylmalonyl-CoA during the breakdown of branched chain amino acids. The gene encoding MCEE maps to human chromosome 2p13.3. Defects in the MCEE gene are the cause of an autosomal

recessive disease known as methylmalonyl-CoA epimerase deficiency (MCEE deficiency), methylmalonyl-CoA racemase deficiency or methylmalonic aciduria type 3, which is characterized by mild to moderate methylmalonic aciduria.

Subcellular Location: Mitochondrion.

## **DISEASE:**

Defects in MCEE are a cause of methylmalonyl-CoA epimerase deficiency (MCEE deficiency) [MIM:251120]; also known as methylmalonyl-CoA racemase deficiency or methylmalonic aciduria type 3. MCEE deficiency is an autosomal recessive inborn error of amino acid metabolism, involving valine, threonine, isoleucine and methionine. This organic aciduria may present in the neonatal period with life-threatening metabolic acidosis, hyperammonemia, feeding difficulties, pancytopenia and coma.

Similarity: joiotech Belongs to the glyoxalase I family.

SWISS: O96PE7

Gene ID: 84693

Database links:

Entrez Gene: 84693 Human

Entrez Gene: 73724 Mouse

Omim: 608419 Human

SwissProt: Q96PE7 Human

SwissProt: Q9D115 Mouse

Unigene: 94949 Human

Unigene: 10093 Mouse

**Important Note:** 

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.