



Rabbit Anti-MUT/Methylmalonyl Coenzyme A mutase antibody

SL18808R

Product Name:	MUT/Methylmalonyl Coenzyme A mutase
Chinese Name:	甲基丙二酰异构酶抗体
Alias:	MCM; Methylmalonyl CoA isomerase; Methylmalonyl CoA mutase mitochondrial; Methylmalonyl Coenzyme A mutase; Methylmalonyl-CoA isomerase; Methylmalonyl-CoA mutase; mitochondrial; Mut; MUTA_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,
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:	79kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human MUT/Methylmalonyl Coenzyme A mutase:451-550/750
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 mitochondrial enzyme methylmalonyl Coenzyme A mutase. In humans, the product of this gene is a vitamin B12-dependent enzyme which catalyzes

the isomerization of methylmalonyl-CoA to succinyl-CoA, while in other species this enzyme may have different functions. Mutations in this gene may lead to various types of methylmalonic aciduria. [provided by RefSeq, Jul 2008]

Function:

Involved in the degradation of several amino acids, odd-chain fatty acids and cholesterol via propionyl-CoA to the tricarboxylic acid cycle. MCM has different functions in other species.

Subcellular Location:

Mitochondrion matrix.

DISEASE:

Defects in MUT are the cause of methylmalonic aciduria type mut (MMAM) [MIM:251000]. MMAM is an often fatal disorder of organic acid metabolism. Common clinical features include lethargy, vomiting, failure to thrive, hypotonia, neurological deficit and early death. Two forms of the disease are distinguished by the presence (mut-) or absence (mut0) of residual enzyme activity. Mut0 patients have more severe neurological manifestations of the disease than do MUT- patients. MMAM is unresponsive to vitamin B12 therapy.

Similarity:

Belongs to the methylmalonyl-CoA mutase family.
Contains 1 B12-binding domain.

SWISS:

P22033

Gene ID:

4594

Database links:

[Entrez Gene: 4594](#) Human

[Entrez Gene: 422049](#) Chicken

[Entrez Gene: 17850](#) Mouse

[Entrez Gene: 688517](#) Rat

[Entrez Gene: 569581](#) Zebrafish

[Omim: 609058](#) Human

[SwissProt: P22033](#) Human

[SwissProt: P16332](#) Mouse

[Unigene: 485527](#) Human

[Unigene: 259884](#) Mouse

Important Note:

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

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