甲基丙二酰异构酶抗体

产品货号： mlR18808

英文名称： MUT/Methylmalonyl Coenzyme A mutase

中文名称： 甲基丙二酰异构酶抗体

别　　名： MCM; Methylmalonyl CoA isomerase; Methylmalonyl CoA mutase mitochondrial; Methylmalonyl Coenzyme A mutase; Methylmalonyl-CoA isomerase; Methylmalonyl-CoA mutase; mitochondrial; Mut; MUTA_HUMAN.

研究领域： 细胞生物　信号转导

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 79kDa

细胞定位： 细胞浆

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原 ： KLH conjugated synthetic peptide derived from human MUT/Methylmalonyl Coenzyme A mutase:451-550/750

亚　　型 ： IgG

纯化方法 ： affinity purified by Protein A

储 存 液 ： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件 ： 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

产品介绍 ： 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

Important Note:

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