甲基丙二酸尿症 B 型蛋白抗体

产品货号： mlR18959

英文名称： MMAB

中文名称： 甲基丙二酸尿症 B 型蛋白抗体

别　　名： aquocob(I)alamin vitamin B12s adenosyltransferase; ATP:cob(I)alamin adenosyltransferase; ATP:corrinoid adenosyltransferase; ATR; c-diamide adenosyltransferase; cblB; Cob; Cob(I)alamin adenosyltransferase; Cob(I)yrinic acid a; cob(I)yrinic acid a c diamide adenosyltransferase mitochondrial; Methylmalonic aciduria (cobalamin deficiency) cblB type; Methylmalonic aciduria type B protein; MGC20496; mitochondrial; MMAB; MMAB gene; MMAB_HUMAN; OTTHUMP00000240563; OTTHUMP00000240564.

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, Chicken, Pig, Cow, Rabbit, Sheep,

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： 24kDa

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 MMAB:101-200/250

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 a protein that catalyzes the final step in the conversion of vitamin B(12) into adenosylcobalamin (AdoCbl), a vitamin B12-containing coenzyme for methylmalonyl-CoA mutase. Mutations in the gene are the cause of vitamin B12-dependent methylmalonic aciduria linked to the cblB complementation group. Alternatively spliced transcript variants have been found. [provided by RefSeq, Apr 2011]

Subcellular Location:

Mitochondrion.

Tissue Specificity:

Expressed in liver and skeletal muscle.

DISEASE:

Defects in MMAB are the cause of methylmalonic aciduria type cblB (MMAB) [MIM:251110]; also known as methylmalonic aciduria type B or vitamin B12-responsive methylmalonicaciduria of cblB complementation type. MMAB is a disorder of methylmalonate and cobalamin metabolism due to defective synthesis of

adenosylcobalamin. Inheritance is autosomal recessive.

Similarity:

Belongs to the Cob(I)alamin adenosyltransferase family.

SWISS:

Q96EY8

Gene ID:

236625

Important Note:

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