δ氨基乙酰丙酸脱水酶抗体

产品货号： mlR7954

英文名称： ALAD

中文名称： δ氨基乙酰丙酸脱水酶抗体

别　名： ALAD; ALADH; ALADR; Aminolevulinate dehydratase; Aminolevulinate, delta, dehydratase; Delta aminolevulinic acid dehydratase; Delta-aminolevulinic acid dehydratase; HEM2_HUMAN; Lv; PBGS; Porphobilinogen synthase.

研究领域： 心血管　细胞生物

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Dog, Cow, Horse, Rabbit, Sheep,

产品应用： WB=1:500-2000 ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 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： 36kDa

细胞定位： 细胞核 细胞浆 细胞外基质 分泌型蛋白

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human ALAD:151-240/330

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 ℃ 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 ℃.

PubMed： PubMed

产品介绍 ： Catalyzes an early step in the biosynthesis of tetrapyrroles. Binds two molecules of 5-aminolevulinate per subunit, each at a distinct site, and catalyzes their condensation to form porphobilinogen. Involvement in disease:Defects in ALAD are the cause of acute hepatic porphyria (AHP). AHP is a form of porphyria. Porphyrias are inherited defects in the biosynthesis of heme, resulting in the accumulation and increased excretion of porphyrins or porphyrin precursors. They are classified as erythropoietic or hepatic, depending on whether the enzyme deficiency occurs in red blood cells or in the liver. AHP is characterized by attacks of gastrointestinal disturbances, abdominal colic, paralysis, and peripheral neuropathy. Most attacks are precipitated by drugs, alcohol, caloric deprivation, infections, or endocrine factors.

Function:

Catalyzes an early step in the biosynthesis of tetrapyrroles. Binds two molecules of 5-aminolevulinate per subunit, each at a distinct site, and catalyzes their condensation to form porphobilinogen.

Subunit:

Homooctamer; active form. Homohexamer; low activity form.

DISEASE:

Defects in ALAD are the cause of acute hepatic porphyria (AHEPP) [MIM:612740]. A form of porphyria. Porphyrias are inherited defects in the biosynthesis of heme, resulting in the accumulation and increased excretion of porphyrins or porphyrin precursors. They are classified as erythropoietic or hepatic, depending on whether the enzyme deficiency occurs in red blood cells or in the liver. AHP is characterized by attacks of gastrointestinal disturbances, abdominal colic, paralysis, and peripheral neuropathy. Most attacks are precipitated by drugs, alcohol, caloric deprivation, infections, or endocrine factors.

Similarity:

Belongs to the ALADH family.

SWISS:

P13716

Gene ID:

210

Important Note:

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