氨基甲酸乙酰转移酶抗体

产品货号： mlR19004

英文名称： NAGS

中文名称： 氨基甲酸乙酰转移酶抗体

别　　名： 1700120E20Rik; AGAS; AI415708; Amino-acid acetyltransferase; ARGA; EC 2.3.1.1; MGC133025; NAGS_HUMAN; N-acetylglutamate synthase; N-acetylglutamate synthase, mitochondrial; RP23-398F7.13.

研究领域： 肿瘤　细胞生物　信号转导　G 蛋白信号

抗体来源： Rabbit

克隆类型： Polyclonal

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

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： 56kDa

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 NAGS:201-300/534

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： The N-acetylglutamate synthase gene encodes a mitochondrial enzyme that catalyzes the formation of N-acetylglutamate (NAG) from glutamate and acetyl coenzyme-A. NAG is a cofactor of carbamyl phosphate synthetase I (CPSI), the first enzyme of the urea cycle in mammals. This gene may regulate ureagenesis by altering NAG availability and, thereby, CPSI activity. Deficiencies in N-acetylglutamate synthase have been associated with hyperammonemia. [provided by RefSeq, Jul 2008]

Function:

Plays a role in the regulation of ureagenesis by producing variable amounts of N-acetylglutamate (NAG), thus modulating carbamoylphosphate synthase I (CPSI) activity.

Subcellular Location:

Mitochondrion matrix.

Tissue Specificity:

Highly expressed in the adult liver, kidney and small intestine. Weakly expressed in the fetal liver, lung, pancreas, placenta, heart and brain tissue.

Post-translational modifications:

Probably processed by mitochondrial processing peptidase (MPP). The long form has not yet been isolated

DISEASE:

N-acetylglutamate synthase deficiency (NAGSD) [MIM:237310]: Rare autosomal recessively inherited metabolic disorder leading to severe neonatal or late-onset hyperammonemia without increased excretion of orotic acid. Clinical symptoms are somnolence, tachypnea, feeding difficulties, a severe neurologic presentation characterized by uncontrollable movements, developmental delay, visual impairment, failure to thrive and hyperammonemia precipitated by the introduction of high-protein diet or febrile illness.

Similarity:

Belongs to the acetyltransferase family.

Contains 1 N-acetyltransferase domain.

SWISS:

Q8N159

Gene ID:

162417

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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Good elisakit producers