

NAGS rabbit pAb

Catalog No: YT6772

Reactivity: Human; Mouse

Applications: WB;IHC

Target: NAGS

Fields: >>Arginine biosynthesis;>>Metabolic pathways;>>2-Oxocarboxylic acid

metabolism;>>Biosynthesis of amino acids

Gene Name: NAGS

Protein Name: NAGS

Human Gene Id: 162417

Human Swiss Prot

No:

Mouse Gene Id: 217214

Mouse Swiss Prot

No:

Immunogen: Synthesized peptide derived from human NAGS AA range: 361-411

Specificity: This antibody detects endogenous levels of NAGS at Human/Mouse

Formulation : Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Source: Polyclonal, Rabbit, IgG

Dilution: WB 1:500-2000;IHC 1:50-300

Q8N159

Q8R4H7

Purification: The antibody was affinity-purified from rabbit antiserum by affinity-

chromatography using epitope-specific immunogen.

Concentration: 1 mg/ml

1/3

Storage Stability: -15°C to -25°C/1 year(Do not lower than -25°C)

Molecularweight: 59kD

Background:

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:

catalytic activity:Acetyl-CoA + L-glutamate = CoA + N-acetyl-L-glutamate.,disease:Defects in NAGS are the cause of N-acetylglutamate synthase deficiency (NAGSD) [MIM:237310]. NAGSD is a 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.,enzyme regulation:Increased by L-arginine.,function:Plays a role in the regulation of ureagenesis by producing variable amounts of N-acetylglutamate (NAG), thus modulating carbamoylphosphate synthase I (CPSI) activity.,online information:N-acetylglutamate synthase

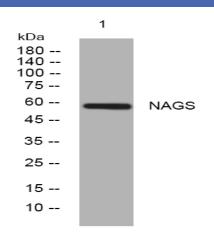
Subcellular Location:

Mitochondrion matrix.

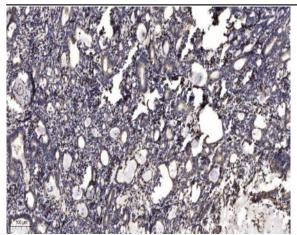
Expression:

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

Products Images



Western blot analysis of lysates from HpeG2 cells, primary antibody was diluted at 1:1000, 4° over night



Immunohistochemical analysis of paraffin-embedded human Gastric adenocarcinoma. 1, Antibody was diluted at 1:200(4° overnight). 2, Tris-EDTA,pH9.0 was used for antigen retrieval. 3,Secondary antibody was diluted at 1:200(room temperature, 45min).