

GAMT Rabbit pAb

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Catalog # AP55119

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q14353
Reactivity	Rat, Human, Mouse
Predicted	Pig, Dog
Host	Rabbit
Clonality	Polyclonal
Calculated MW	26318
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human GAMT
Epitope Specificity	101-200/236
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
DISEASE	Defects in GAMT are the cause of guanidinoacetate methyltransferase deficiency (GAMT deficiency) [MIM:612736]. GAMT deficiency is an autosomal recessive disorder characterized by developmental delay/regression, mental retardation, severe disturbance of expressive and cognitive speech, intractable seizures and movement disturbances, severe depletion of creatine/phosphocreatine in the brain, and accumulation of guanidinoacetic acid (GAA) in brain and body fluids.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	In the creatine biosynthesis pathway, glycine is converted to guanidinoacetate by amidinotransferase, and guanidinoacetate is then converted to creatine by Guanidinoacetate N-methyltransferase (GAMT). GAMT, a methyltransferase, uses S-adenosylmethionine as the methyl donor for this reaction. Methyltransferases are a type of transferase enzyme which transfers a methyl group to nucleic bases in DNA or amino acids in protein. Encoding a 236 amino acid protein, the human GAMT gene maps to chromosome 19p13.3. Defects in the GAMT gene leads to GAMT deficiency, which is associated with guanidinoacetate accumulation and decreased levels of creatine excretion in brain. Such biochemical changes are thought to lead to various neurological syndromes and muscular hypotonia.

Additional Information

Gene ID	2593
Other Names	Guanidinoacetate N-methyltransferase, 2.1.1.2, GAMT
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500

Storage Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name GAMT

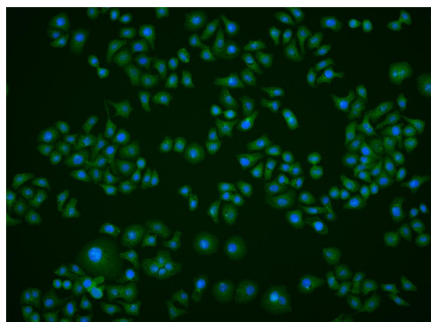
Function Converts guanidinoacetate to creatine, using S- adenosylmethionine as the methyl donor (PubMed:[24415674](#), PubMed:[26003046](#), PubMed:[26319512](#)). Important in nervous system development (PubMed:[24415674](#)).

Tissue Location Expressed in liver..

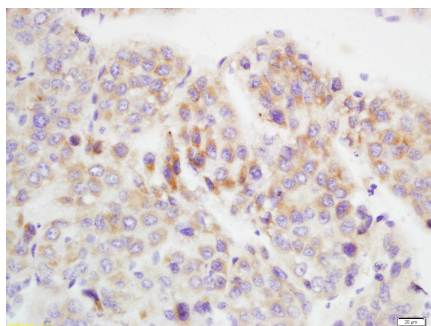
Background

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Images



HepG2 cell; 4% Paraformaldehyde-fixed; Triton X-100 at room temperature for 20 min; Blocking buffer (normal goat serum, C-0005) at 37°C for 20 min; Antibody incubation with (GAMT) polyclonal Antibody, Unconjugated (AP55119) 1:50, 90 minutes at 37°C; followed by a conjugated Goat Anti-Rabbit IgG antibody at 37°C for 90 minutes, DAPI (blue, C02-04002) was used to stain the cell nuclei.



Tissue/cell: Human hepatocellular carcinoma; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37°C for 20 min; Incubation: Anti-GMAT Polyclonal Antibody, Unconjugated(AP55119) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.