10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

GATM Antibody(C-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP19715b

Product Information

Application	WB, E
Primary Accession	P50440
Other Accession	P50442, P50441, Q9D964, Q4R806, Q2HJ74, NP_001473.1
Reactivity	Human
Predicted	Bovine, Monkey, Mouse, Pig, Rat
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB41023
Calculated MW	48455
Antigen Region	350-378

Additional Information

Gene ID	2628
Other Names	Glycine amidinotransferase, mitochondrial, L-arginine:glycine amidinotransferase, Transamidinase, GATM, AGAT
Target/Specificity	This GATM antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 350-378 amino acids from the C-terminal region of human GATM.
Dilution	WB~~1:1000 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	GATM Antibody(C-term) is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	GATM
Synonyms	AGAT

Function	Transamidinase that catalyzes the transfer of the amidino group of L-arginine onto the amino moiety of acceptor metabolites such as glycine, beta-alanine, gamma-aminobutyric acid (GABA) and taurine yielding the corresponding guanidine derivatives (PubMed:16820567, PubMed:27233232, PubMed:36543883, PubMed:3800397). Catalyzes the rate- limiting step of creatine biosynthesis, namely the transfer of the amidino group from L-arginine to glycine to generate guanidinoacetate, which is then methylated by GAMT to form creatine. Provides creatine as a source for ATP generation in tissues with high energy demands, in particular skeletal muscle, heart and brain (Probable) (PubMed:27233232, PubMed:36543883, PubMed:3800397, PubMed:9266688).
Cellular Location	[Isoform 1]: Mitochondrion inner membrane; Peripheral membrane protein; Intermembrane side. Note=Probably attached to the outer side of the inner membrane
Tissue Location	Expressed in brain, heart, kidney, liver, lung, salivary gland and skeletal muscle tissue, with the highest expression in kidney. Biallelically expressed in placenta and fetal tissues

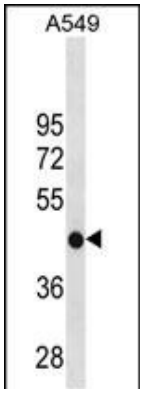
Background

This gene encodes a mitochondrial enzyme that belongs to the amidinotransferase family. This enzyme is involved in creatine biosynthesis, whereby it catalyzes the transfer of a guanido group from L-arginine to glycine, resulting in guanidinoacetic acid, the immediate precursor of creatine. Mutations in this gene cause arginine:glycine amidinotransferase deficiency, an inborn error of creatine synthesis characterized by mental retardation, language impairment, and behavioral disorders.

References

Kottgen, A., et al. Nat. Genet. 42(5):376-384(2010)
Kottgen, A., et al. Nat. Genet. 41(6):712-717(2009)
Lion-Francois, L., et al. Neurology 67(9):1713-1714(2006)
Cullen, M.E., et al. Circulation 114 (1 SUPPL), I16-I20 (2006) :
Battini, R., et al. J. Pediatr. 148(6):828-830(2006)

Images



GATM Antibody (C-term) (Cat. #AP19715b) western blot analysis in A549 cell line lysates (35ug/lane).This demonstrates the GATM antibody detected the GATM protein (arrow).