SANTA CRUZ BIOTECHNOLOGY, INC.

GAMT (A-11): sc-398936

BACKGROUND

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.

REFERENCES

1. Stöckler, S., et al. 1994. Creatine deficiency in the brain: a new, treatable inborn error of metabolism. Pediatr. Res. 36: 409-413.
2. Isbrandt, D. and von Figura, K. 1995. Cloning and sequence analysis of human guanidinoacetate N-methyltransferase cDNA. Biochim. Biophys. Acta 1264: 265-267.
3. Jenne, D.E., et al. 1997. The human guanidinoacetate methyltransferase (GAMT) gene maps to a syntenic region on 19p13.3, homologous to band C of mouse chromosome 10, but GAMT is not mutated in jittery mice. Biochem. Biophys. Res. Commun. 238: 723-727.
4. Schulze, A., et al. 1997. Creatine deficiency syndrome caused by guanidinoacetate methyltransferase deficiency: diagnostic tools for a new inborn error of metabolism. J. Pediatr. 131: 626-631.
5. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 601240. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/
6. Wang, Y., et al. 2006. Tyrosine phosphorylated PAR3 regulates epithelial tight junction assembly promoted by EGFR signaling. EMBO J. 25: 5058-5070.
7. Almeida, L.S., et al. 2006. Overexpression of GAMT restores GAMT activity in primary GAMT-deficient fibroblasts. Mol. Genet. Metab. 89: 392-394.
8. Morris, A.A., et al. 2007. Guanidinoacetate methyltransferase deficiency masquerading as a mitochondrial encephalopathy. J. Inherit. Metab. Dis. 30: 100.

CHROMOSOMAL LOCATION

Genetic locus: GAMT (human) mapping to 19p13.3.

SOURCE

GAMT (A-11) is a mouse monoclonal antibody raised against amino acids 1-160 mapping at the N-terminus of GAMT of human origin.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

PRODUCT

Each vial contains 200 µg IgG_1 in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

GAMT (A-11) is available conjugated to agarose (sc-398936 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-398936 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-398936 PE), fluorescein (sc-398936 FITC), Alexa Fluor® 488 (sc-398936 AF488), Alexa Fluor® 546 (sc-398936 AF546), Alexa Fluor® 594 (sc-398936 AF594) or Alexa Fluor® 647 (sc-398936 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-398936 AF680) or Alexa Fluor® 790 (sc-398936 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

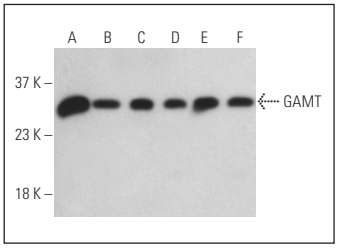
GAMT (A-11) is recommended for detection of GAMT of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for GAMT siRNA (h): sc-97156, GAMT shRNA Plasmid (h): sc-97156-SH and GAMT shRNA (h) Lentiviral Particles: sc-97156-V.

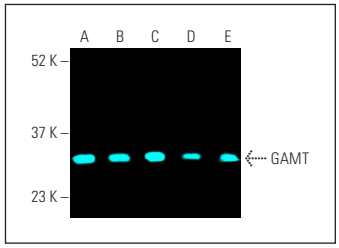
Molecular Weight of GAMT: 26 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, Jurkat whole cell lysate: sc-2204 or SH-SY5Y cell lysate: sc-3812.

DATA



GAMT (A-11): sc-398936. Western blot analysis of GAMT expression in Hep G2 (**A**), Daoy (**B**), Jurkat (**C**), SH-SY5Y (**D**), MOLT-4 (**E**) and RPMI-8226 (**F**) whole cell lysates. Detection reagent used: m-IgG_1 BP-HRP: sc-525408.



GAMT (A-11): sc-398936. Fluorescent western blot analysis of GAMT expression in MOLT-4 (**A**), Hep G2 (**B**), Jurkat (**C**), Daoy (**D**) and RPMI-8226 (**E**) whole cell lysates. Blocked with UltraCruz® Blocking Reagent: sc-516214. Detection reagent used: m-IgG_1 BP-CFL 647: sc-533664.

SELECT PRODUCT CITATIONS

1. Chen, H.R., et al. 2021. Creatine transporter deficiency impairs stress adaptation and brain energetics homeostasis. JCI Insight 6: 140173.

RESEARCH USE

For research use only, not for use in diagnostic procedures.