SANTA CRUZ BIOTECHNOLOGY, INC.

NAGS (E-8): sc-515127

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

The function of the urea cycle is to remove excess nitrogen from the body. Six distinct enzymes comprise the urea cycle, and urea cycle disorders (UCD) are a direct results of deficiency in any one of those enzymes. N-acetylglutamate synthase (NAGS) catalyzes the conversion of N-acetylglutamate (NAG) from glutamate and acetylcoenzyme A. NAG is an obligatory activator of carbamylphosphate I (CPSI), the first and rate limiting enzyme of ureagenesis. Therefore, deficiency of NAGS results in severe hyperammonemia. Twenty one mutations have been described in humans, ten of which are associated with acute neonatal hyperammonemia, and the remainder found in patients with late onset disease. Treatment with N-carbamylglutamate (NCG) can ameliorate hyperammonemia for inherited and secondary NAGS deficiency. Expression of NAGS occurs in the liver, small intestine and kidney.

REFERENCES

1. Caldovic, L., et al. 2002. Cloning and expression of the human N-acetylglutamate synthase gene. Biochem. Biophys. Res. Commun. 299: 581-586.
2. Häberle, J., et al. 2003. Mutation analysis in patients with N-acetylglutamate synthase deficiency. Hum. Mutat. 21: 593-597.
3. Morizono, H., et al. 2004. Mammalian N-acetylglutamate synthase. Mol. Genet. Metab. 81: S4-S11.
4. Caldovic, L., et al. 2006. Biochemical properties of recombinant human and mouse N-acetylglutamate synthase. Mol. Genet. Metab. 87: 226-232.
5. Caldovic, L., et al. 2007. Mutations and polymorphisms in the human N-acetylglutamate synthase (NAGS) gene. Hum. Mutat. 28: 754-759.
6. Nordenström, A., et al. 2007. A trial with N-carbamylglutamate may not detect all patients with NAGS deficiency and neonatal onset. J. Inherit. Metab. Dis. 30: 400.

CHROMOSOMAL LOCATION

Genetic locus: NAGS (human) mapping to 17q21.31.

SOURCE

NAGS (E-8) is a mouse monoclonal antibody raised against amino acids 226-341 mapping within an internal region of NAGS of human origin.

PRODUCT

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

NAGS (E-8) is available conjugated to agarose (sc-515127 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-515127 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-515127 PE), fluorescein (sc-515127 FITC), Alexa Fluor® 488 (sc-515127 AF488), Alexa Fluor® 546 (sc-515127 AF546), Alexa Fluor® 594 (sc-515127 AF594) or Alexa Fluor® 647 (sc-515127 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-515127 AF680) or Alexa Fluor® 790 (sc-515127 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

NAGS (E-8) is recommended for detection of NAGS 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 NAGS siRNA (h): sc-93810, NAGS shRNA Plasmid (h): sc-93810-SH and NAGS shRNA (h) Lentiviral Particles: sc-93810-V.

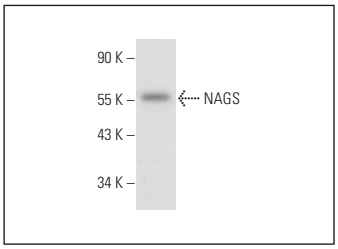
Molecular Weight of NAGS: 58 kDa.

Positive Controls: human liver extract: sc-363766.

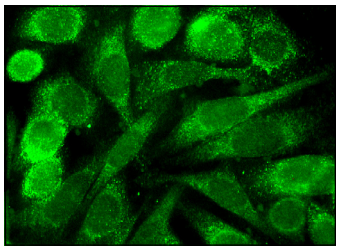
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



NAGS (E-8): sc-515127. Western blot analysis of NAGS expression in human liver tissue extract.



NAGS (E-8): sc-515127. Immunofluorescence staining of formalin-fixed SW480 cells showing mitochondrial localization.

STORAGE

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

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.