

# NAGS (Q-12): sc-132384

## 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. Inher. Metab. Dis.* 30: 400.
7. Deignan, J.L., et al. 2008. Contrasting features of urea cycle disorders in human patients and knockout mouse models. *Mol. Genet. Metab.* 93: 7-14.

## CHROMOSOMAL LOCATION

Genetic locus: NAGS (human) mapping to 17q21.31; Nags (mouse) mapping to 11 D.

## SOURCE

NAGS (Q-12) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of NAGS of human origin.

## PRODUCT

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

Blocking peptide available for competition studies, sc-132384 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## STORAGE

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

## APPLICATIONS

NAGS (Q-12) is recommended for detection of NAGS of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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).

NAGS (Q-12) is also recommended for detection of NAGS in additional species, including equine, canine and bovine.

Suitable for use as control antibody for NAGS siRNA (h): sc-93810, NAGS siRNA (m): sc-149805, NAGS shRNA Plasmid (h): sc-93810-SH, NAGS shRNA Plasmid (m): sc-149805-SH, NAGS shRNA (h) Lentiviral Particles: sc-93810-V and NAGS shRNA (m) Lentiviral Particles: sc-149805-V.

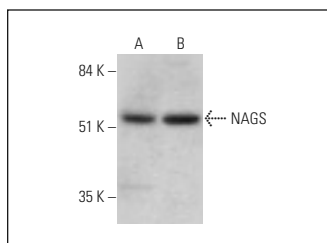
Molecular Weight of NAGS: 58 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, human liver extract: sc-363766 or mouse small intestine extract: sc-364252.

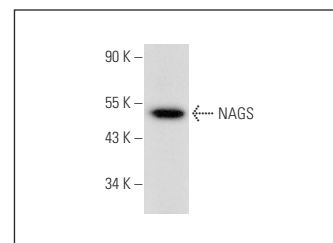
## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## DATA



NAGS (Q-12): sc-132384. Western blot analysis of NAGS expression in Hep G2 whole cell lysate (A) and human liver tissue extract (B).



NAGS (Q-12): sc-132384. Western blot analysis of NAGS expression in mouse small intestine tissue extract.

## RESEARCH USE

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

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Try **NAGS (E-8): sc-515127**, our highly recommended monoclonal alternative to NAGS (Q-12).