

## GLCNE (H-300): sc-134809

### BACKGROUND

The bifunctional enzyme UDP-N-acetylglucosamine 2-epimerase/N-acetylmannosamine kinase (GNE/Mnk), or GLCNE, regulates and initiates biosynthesis of N-acetylneuraminic acid (NeuAc), a precursor of sialic acids. GLCNE is required for normal sialylation in hematopoietic cells. Sialylation is implicated in cell adhesion, signal transduction, tumorigenicity and metastatic behavior of malignant cells. It is upregulated after PKC-dependent phosphorylation and is most abundantly expressed in liver and placenta. It is also expressed, to a lesser extent, in heart, brain, lung, kidney, skeletal muscle and pancreas. Defects in GLCNE are the cause of sialuria, inclusion body myopathy 2 (IBM2) and Nonaka myopathy (NM) or distal myopathy with rimmed vacuoles (DMRV). Sialuria is an autosomal dominant disorder caused by a lack of feedback inhibition of GLCNE by CMP-NeuAc, resulting in overproduction of NeuAc. It is characterized by an accumulation of free sialic acid in the cytoplasm and large quantities of neuraminic acid in the urine. Both IBM2 and NM/DMRV are autosomal recessive neuromuscular disorders characterized by adult onset, distal and proximal muscle weakness (especially in the legs) and a typical muscle pathology including filamentous inclusions and rimmed vacuoles.

### REFERENCES

1. Amouri, R., et al. 2005. Allelic heterogeneity of GNE gene mutation in two Tunisian families with autosomal recessive inclusion body myopathy. *Neuromuscul. Disord.* 15: 361-363.
2. Bork, K., et al. 2005. The intracellular concentration of sialic acid regulates the polysialylation of the neural cell adhesion molecule. *FEBS Lett.* 579: 5079-5083.
3. Krause, S., et al. 2005. Localization of UDP-GlcNAc 2-epimerase/ManAc kinase (GNE) in the Golgi complex and the nucleus of mammalian cells. *Exp. Cell Res.* 304: 365-379.
4. Nonaka, I., et al. 2005. Distal myopathy with rimmed vacuoles and hereditary inclusion body myopathy. *Curr. Neurol. Neurosci. Rep.* 5: 61-65.
5. Ro, L.S., et al. 2005. Phenotypic variability in a Chinese family with rimmed vacuolar distal myopathy. *J. Neurol. Neurosurg. Psychiatry* 76: 752-755.

### CHROMOSOMAL LOCATION

Genetic locus: GNE (human) mapping to 9p13.3; Gne (mouse) mapping to 4 B1.

### SOURCE

GLCNE (H-300) is a rabbit polyclonal antibody raised against amino acids 1-300 mapping at the N-terminus of GLCNE 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.

### RESEARCH USE

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

### APPLICATIONS

GLCNE (H-300) is recommended for detection of GLCNE 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).

GLCNE (H-300) is also recommended for detection of GLCNE in additional species, including equine, canine, bovine, porcine and avian.

Suitable for use as control antibody for GLCNE siRNA (h): sc-60693, GLCNE siRNA (m): sc-60694, GLCNE shRNA Plasmid (h): sc-60693-SH, GLCNE shRNA Plasmid (m): sc-60694-SH, GLCNE shRNA (h) Lentiviral Particles: sc-60693-V and GLCNE shRNA (m) Lentiviral Particles: sc-60694-V.

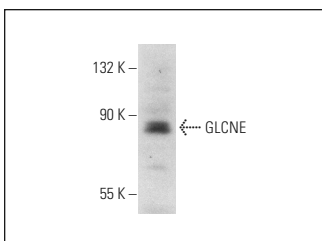
Molecular Weight of GLCNE: 79 kDa.

Positive Controls: mouse brain extract: sc-2253.

### RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

### DATA



GLCNE (H-300): sc-134809. Western blot analysis of GLCNE expression in mouse brain tissue extract.

### STORAGE

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

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Try **GLCNE (H-10): sc-376057** or **GLCNE (D-8): sc-374512**, our highly recommended monoclonal alternatives to GLCNE (H-300).