SANTA CRUZ BIOTECHNOLOGY, INC.

GLCNE (D-8): sc-374512

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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. Psychiatr. 76: 752-755.
6. Salama, I., et al. 2005. No overall hyposialylation in hereditary inclusion body myopathy myoblasts carrying the homozygous M712T GNE mutation. Biochem. Biophys. Res. Commun. 328: 221-226.
7. Sparks, S.E., et al. 2005. Use of a cell-free system to determine UDP-N-acetylglucosamine 2-epimerase and N-acetylmannosamine kinase activities in human hereditary inclusion body myopathy. Glycobiology 15: 1102-1110.

CHROMOSOMAL LOCATION

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

SOURCE

GLCNE (D-8) is a mouse monoclonal antibody raised against amino acids 1-300 mapping at the N-terminus of GLCNE of human origin.

RESEARCH USE

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

PRODUCT

Each vial contains 200 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

GLCNE (D-8) is recommended for detection of GLCNE of mouse, rat and 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).

GLCNE (D-8) is also recommended for detection of GLCNE in additional species, including equine, canine and porcine.

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.

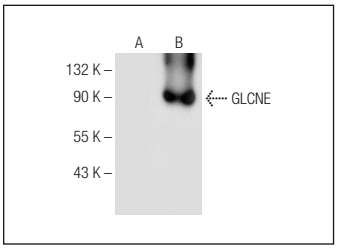
Molecular Weight of GLCNE: 79 kDa.

Positive Controls: GLCNE (m2): 293T Lysate: sc-125387 or human liver extract: sc-363766.

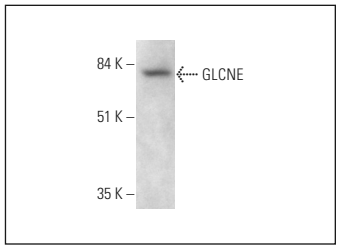
RECOMMENDED SUPPORT REAGENTS

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



GLCNE (D-8): sc-374512. Western blot analysis of GLCNE expression in non-transfected: sc-117752 (**A**) and mouse GLCNE transfected: sc-125387 (**B**) 293T whole cell lysates.



GLCNE (D-8): sc-374512. Western blot analysis of GLCNE expression in human liver tissue extract.

STORAGE

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