SANTA CRUZ BIOTECHNOLOGY, INC.

GLCNE (H-10): sc-376057

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.

CHROMOSOMAL LOCATION

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

SOURCE

GLCNE (H-10) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 48-77 at the N-terminus of GLCNE of human origin.

PRODUCT

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

GLCNE (H-10) is available conjugated to agarose (sc-376057 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-376057 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-376057 PE), fluorescein (sc-376057 FITC), Alexa Fluor® 488 (sc-376057 AF488), Alexa Fluor® 546 (sc-376057 AF546), Alexa Fluor® 594 (sc-376057 AF594) or Alexa Fluor® 647 (sc-376057 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-376057 AF680) or Alexa Fluor® 790 (sc-376057 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-376057 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

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APPLICATIONS

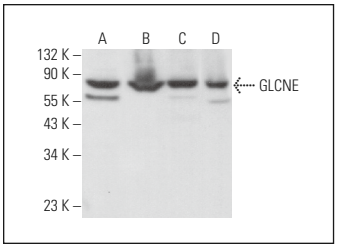
GLCNE (H-10) is recommended for detection of GLCNE of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

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.

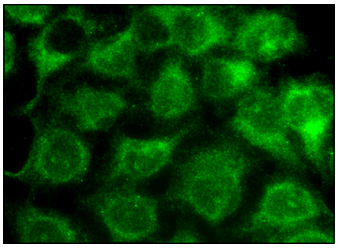
Molecular Weight of GLCNE: 79 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, c4 whole cell lysate: sc-364186 or Caco-2 cell lysate: sc-2262.

DATA



GLCNE (H-10): sc-376057. Western blot analysis of GLCNE expression in Hep G2 (**A**), c4 (**B**), Caco-2 (**C**) and PC-12 (**D**) whole cell lysates.



GLCNE (H-10): sc-376057. Immunofluorescence staining of methanol-fixed HeLa cells showing cytoplasmic localization.

SELECT PRODUCT CITATIONS

1. Devi, S.S., et al. 2021. Generation and characterization of a skeletal muscle cell-based model carrying one single Gne allele: implications in Actin dynamics. Mol. Neurobiol. 58: 6316-6334.
2. Bottega, R., et al. 2022. GNE-related thrombocytopenia: evidence for a mutational hotspot in the ADP/substrate domain of the GNE bifunctional enzyme. Haematologica 107: 750-754.
3. Sharma, S., et al. 2022. Functional characterization of GNE mutations prevalent in Asian subjects with GNE myopathy, an ultra-rare neuromuscular disorder. Biochimie 199: 36-45.
4. Neu, C.T., et al. 2024. GNE deficiency impairs Myogenesis in C2C12 cells and cannot be rescued by ManNAc supplementation. Glycobiology 34: cwae004.

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.