SANTA CRUZ BIOTECHNOLOGY, INC.

GLDC (A-9): sc-376106

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

The glycine cleavage system is comprised of AMT (known as protein T), GCSH (known as protein H), DLD (known as protein L) and GLDC (known as protein P), all of which work together to catalyze the cleavage and degradation of glycine. GLDC (glycine dehydrogenase), also known as GCE, GCSP (glycine cleavage system P protein) or HYGN1, is a 1,020 amino acid protein that localizes to the mitochondria and belongs to the gcvP family. GLDC binds to glycine and enables the methylamine group from glycine to be transferred to the protein T. GLDC exists as a homodimer and utilizes pyridoxal phosphate as a cofactor. Mutations in the gene encoding GLDC leads to nonketotic hyperglycinemia (NKH), also known as glycine encephalopathy (GCE), an autosomal recessive disease characterized by accumulation of a large amount of glycine in body fluid and by severe neurological symptoms.

REFERENCES

1. Takayanagi, M., et al. 2000. Human glycine decarboxylase gene (GLDC) and its highly conserved processed pseudogene (psiGLDC): their structure and expression, and the identification of a large deletion in a family with non-ketotic hyperglycinemia. Hum. Genet. 106: 298-305.
2. Kure, S., et al. 2002. Heterozygous GLDC and GCSH gene mutations in transient neonatal hyperglycinemia. Ann. Neurol. 52: 643-646.
3. Toone, J.R., et al. 2002. Novel mutations in the P-protein (glycine decarboxylase) gene in patients with glycine encephalopathy (non-ketotic hyperglycinemia). Mol. Genet. Metab. 76: 243-249.
4. Flusser, H., et al. 2005. Mild glycine encephalopathy (NKH) in a large kindred due to a silent exonic GLDC splice mutation. Neurology 64: 1426-1430.
5. Conter, C., et al. 2006. Genetic heterogeneity of the GLDC gene in 28 unrelated patients with glycine encephalopathy. J. Inherit. Metab. Dis. 29: 135-142.
6. Kanno, J., et al. 2007. Genomic deletion within GLDC is a major cause of non-ketotic hyperglycinaemia. J. Med. Genet. 44: e69.
7. Oda, M., et al. 2007. Direct correlation between ischemic injury and extracellular glycine concentration in mice with genetically altered activities of the glycine cleavage multienzyme system. Stroke 38: 2157-2164.
8. Chang, C.Y., et al. 2008. Non-ketotic hyperglycinemia with a novel GLDC mutation in a Taiwanese child. Acta Paediatr. Taiwan. 49: 35-37.
9. Hellani, A., et al. 2008. Delivery of a normal baby after preimplantation genetic diagnosis for non-ketotic hyperglycinaemia. Reprod. Biomed. Online 16: 893-897.

CHROMOSOMAL LOCATION

Genetic locus: GLDC (human) mapping to 9p24.1; Gldc (mouse) mapping to 19 C1.

SOURCE

GLDC (A-9) is a mouse monoclonal antibody raised against amino acids 256-555 mapping within an internal region of GLDC 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.

APPLICATIONS

GLDC (A-9) is recommended for detection of GLDC 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 GLDC siRNA (h): sc-92873, GLDC siRNA (m): sc-145419, GLDC shRNA Plasmid (h): sc-92873-SH, GLDC shRNA Plasmid (m): sc-145419-SH, GLDC shRNA (h) Lentiviral Particles: sc-92873-V and GLDC shRNA (m) Lentiviral Particles: sc-145419-V.

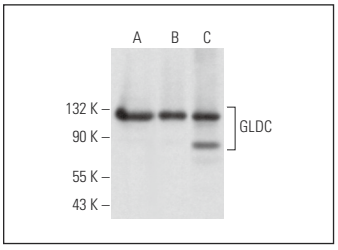
Molecular Weight of GLDC: 113 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, Caco-2 cell lysate: sc-2262 or rat liver extract: sc-2395.

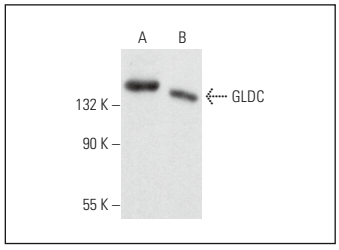
RECOMMENDED SUPPORT REAGENTS

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



GLDC (A-9): sc-376106. Western blot analysis of GLDC expression in Hep G2 (**A**) and Caco-2 (**B**) whole cell lysates and rat liver tissue extract (**C**).



GLDC (A-9): sc-376106. Western blot analysis of GLDC expression in Hep G2 (**A**) and ACHN (**B**) whole cell lysates.

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.