SANTA CRUZ BIOTECHNOLOGY, INC.

GLDC (I-14): sc-162872

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. 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 (I-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of GLDC of human origin.

STORAGE

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

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-162872 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

GLDC (I-14) is recommended for detection of GLDC of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

GLDC (I-14) is also recommended for detection of GLDC in additional species, including equine, canine, bovine, porcine and avian.

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 or mouse liver extract: sc-2256.

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) 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.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.