BACKGROUND

GCSH (glycine cleavage system protein H (aminomethyl carrier)), also known as GCE or NKH, is a 173 amino acid mitochondrial protein that contains one lipoyl-binding domain and belongs to the gcVH family. Defects in the gene encoding GCSH are the cause of glycine encephalopathy (GCE), an autosomal recessive disease that is also referred to as non-ketotic hyperglycinemia (NKH). Characterized by severe neurological symptoms, patients with GCE have a large amount of glycine accumulated in their body fluids. The gene encoding GCSH maps to human chromosome 16, which encodes over 900 genes and comprises nearly 3% of the human genome.

REFERENCES


CHROMOSOMAL LOCATION

Genetic locus: GCSH (human) mapping to 16q23.2; Gcsh (mouse) mapping to 8 E1.

SOURCE

GCSH (S-16) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of GCSH 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-242901 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

GCSH (S-16) is recommended for detection of GCSH 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).

GCSH (S-16) is also recommended for detection of GCSH in additional species, including equine, canine, bovine and avian.

Suitable for use as control antibody for GCSH siRNA (h): sc-93301, GCSH siRNA (m): sc-145367, GCSH shRNA Plasmid (h): sc-93301-SH, GCSH shRNA Plasmid (m): sc-145367-SH, GCSH shRNA (h) Lentiviral Particles: sc-93301-V and GCSH shRNA (m) Lentiviral Particles: sc-145367-V.

Molecular Weight of GCSH: 19 kDa.

Positive Controls: mouse placenta extract: sc-364247 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) Immunoprecipitation: use Protein A/G PLUS-agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) 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.

DATA

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RESEARCH USE

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

GCSH (S-16): sc-242901. Western blot analysis of GCSH expression in mouse liver (A) and mouse placenta (B) tissue extracts.