

PHKG1 (N-12): sc-138526

BACKGROUND

PHKG1 (Phosphorylase kinase subunit γ 1), is a subunit of phosphorylase kinase (PHK) that belongs to the Ser/Thr protein kinase family. PHK is a hexadecameric protein composed of four α chains, four β chains, four γ chains and four δ chains. The γ chains are catalytic chains, the α and β chains are regulatory chains and the δ chains are calmodulins. PHKG1 contains two calmodulin-binding domains and one protein kinase domain. As the catalytic chain of PHK, PHKG1 is responsible for catalyzing the phosphorylation and activation of glycogen phosphorylase and therefore it plays an important role in the glycogenolytic pathway. Mutations in the gene encoding PHKG1 can lead to PHK deficiency and result in glycogen storage disease type 9C (GSD9C), also known as autosomal liver glycogenesis.

REFERENCES

- Hanks, S.K. 1989. Messenger ribonucleic acid encoding an apparent isoform of phosphorylase kinase catalytic subunit is abundant in the adult testis. *Mol. Endocrinol.* 3: 110-116.
- Calalb, M.B., et al. 1992. Molecular cloning and enzymatic analysis of the rat homolog of "PhK-gamma T," an isoform of phosphorylase kinase catalytic subunit. *J. Biol. Chem.* 267: 1455-1463.
- Liu, L., et al. 1996. The testis isoform of the phosphorylase kinase catalytic subunit (PhK- γ T) plays a critical role in regulation of glycogen mobilization in developing lung. *J. Biol. Chem.* 271: 11761-11766.
- Maichele, A.J., et al. 1996. Mutations in the testis/liver isoform of the phosphorylase kinase gamma subunit (PHKG2) cause autosomal liver glycogenesis in the *gsd* rat and in humans. *Nat. Genet.* 14: 337-340.
- Burwinkel, B., et al. 1998. Liver glycogenesis due to phosphorylase kinase deficiency: PHKG2 gene structure and mutations associated with cirrhosis. *Hum. Mol. Genet.* 7: 149-154.
- Burwinkel, B., et al. 2000. Phosphorylase kinase deficient liver glycogenesis: progression to cirrhosis in infancy associated with PHKG2 mutations (H14 and L225R). *J. Med. Genet.* 37: 376-377.
- Burwinkel, B., et al. 2003. Severe phenotype of phosphorylase kinase-deficient liver glycogenesis with mutations in the PHKG2 gene. *Pediatr. Res.* 54: 834-839.
- Chen, C.S., et al. 2006. Effects of *Scutellariae Radix* on gene expression in HEK 293 cells using cDNA microarray. *J. Ethnopharmacol.* 105: 346-351.
- Beauchamp, N.J., et al. 2007. Glycogen storage disease type IX: High variability in clinical phenotype. *Mol. Genet. Metab.* 92: 88-99.

CHROMOSOMAL LOCATION

Genetic locus: PHKG1 (human) mapping to 7p11.2; Phkg1 (mouse) mapping to 5 G1.3.

SOURCE

PHKG1 (N-12) is an affinity purified rabbit polyclonal antibody raised against a peptide mapping near the N-terminus of PHKG1 of human origin.

PRODUCT

Each vial contains 100 μ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-138526 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

PHKG1 (N-12) is recommended for detection of PHKG1 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:50-1:500), immunofluorescence (starting dilution 1:25, dilution range 1:25-1:250) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000); non cross-reactive with PHKG2.

PHKG1 (N-12) is also recommended for detection of PHKG1 in additional species, including bovine and porcine.

Suitable for use as control antibody for PHKG1 siRNA (h): sc-89501, PHKG1 siRNA (m): sc-152225, PHKG1 shRNA Plasmid (h): sc-89501-SH, PHKG1 shRNA Plasmid (m): sc-152225-SH, PHKG1 shRNA (h) Lentiviral Particles: sc-89501-V and PHKG1 shRNA (m) Lentiviral Particles: sc-152225-V.

Molecular Weight of PHKG1: 45 kDa.

Positive Controls: Ramos cell lysate: sc-2216.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

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.

PROTOCOLS

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