

PHKG2 (H-68): sc-292918

BACKGROUND

PHKG2 (phosphorylase kinase subunit γ 2), also known as PHK γ testis/liver isoform (PHK- γ -T) or PSK-C3, is a subunit of phosphorylase kinase (PHK). 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. PHKG2 is an isozyme of the γ -chain and is expressed in testis, liver and possibly other non-muscle tissues. It contains one protein kinase domain and belongs to the Ser/Thr protein kinase family. As the catalytic chain of PHK, PHKG2 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 PHKG2 can lead to PHK deficiency and result in glycogen storage disease type 9C (GSD9C), also known as autosomal liver glycogenosis.

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., Fox, D.T. and Hanks, S.K. 1992. Molecular cloning and enzymatic analysis of the rat homolog of "PhK- γ T," an isoform of phosphorylase kinase catalytic subunit. *J. Biol. Chem.* 267: 1455-1463.
- Liu, L., Rannels, S.R., Falconieri, M., Phillips, K.S., Wolpert, E.B. and Weaver, T.E. 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., Burwinkel, B., Maire, I., Sovik, O. and Kilimann, M.W. 1996. Mutations in the testis/liver isoform of the phosphorylase kinase γ subunit (PHKG2) cause autosomal liver glycogenosis in the *gsd* rat and in humans. *Nat. Genet.* 14: 337-340.
- Burwinkel, B., Shiomi, S., Al Zaben, A. and Kilimann, M.W. 1998. Liver glycogenosis due to phosphorylase kinase deficiency: PHKG2 gene structure and mutations associated with cirrhosis. *Hum. Mol. Genet.* 7: 149-154.
- Burwinkel, B., Tanner, M.S. and Kilimann, M.W. 2000. Phosphorylase kinase deficient liver glycogenosis: progression to cirrhosis in infancy associated with PHKG2 mutations (H144Y and L225R). *J. Med. Genet.* 37: 376-377.
- Burwinkel, B., Rootwelt, T., Kvittingen, E.A., Chakraborty, P.K. and Kilimann, M.W. 2003. Severe phenotype of phosphorylase kinase-deficient liver glycogenosis with mutations in the PHKG2 gene. *Pediatr. Res.* 54: 834-839.
- Chen, C.S., Chen, N.J., Lin, L.W., Hsieh, C.C., Chen, G.W. and Hsieh, M.T. 2006. Effects of *Scutellariae Radix* on gene expression in HEK 293 cells using cDNA microarray. *J. Ethnopharmacol.* 105: 346-351.
- Beauchamp, N.J., Dalton, A., Ramaswami, U., Niinikoski, H., Mention, K., Kenny, P., Kolho, K.L., Raiman, J., Walter, J., Treacy, E., Tanner, S. and Sharrard, M. 2007. Glycogen storage disease type IX: High variability in clinical phenotype. *Mol. Genet. Metab.* 92: 88-99.

RESEARCH USE

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

CHROMOSOMAL LOCATION

Genetic locus: PHKG2 (human) mapping to 16p11.2; Phkg2 (mouse) mapping to 7 F3.

SOURCE

PHKG2 (H-68) is a rabbit polyclonal antibody raised against amino acids 39-106 mapping near the N-terminus of PHKG2 of human origin.

PRODUCT

Each vial contains 200 μ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

PHKG2 (H-68) is recommended for detection of PHKG2 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g 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); non cross-reactive with other PHKG family members.

PHKG2 (H-68) is also recommended for detection of PHKG2 in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for PHKG2 siRNA (h): sc-72305, PHKG2 siRNA (m): sc-72306, PHKG2 shRNA Plasmid (h): sc-72305-SH, PHKG2 shRNA Plasmid (m): sc-72306-SH, PHKG2 shRNA (h) Lentiviral Particles: sc-72305-V and PHKG2 shRNA (m) Lentiviral Particles: sc-72306-V.

Molecular Weight of PHKG2: 41 kDa.

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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) 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.

PROTOCOLS

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