SANTA CRUZ BIOTECHNOLOGY, INC.

PHKG1 (RR-34): sc-100536

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The Power to Question

BACKGROUND

PHKG1 (phosphorylase kinase subunit γ1), 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 glycogenosis.

REFERENCES

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3. Liu, L., et al. 1996. The testis isoform of the phosphorylase kinase catalytic subunit (PhK-γT) plays a critical role in regulation of glycogen mobilization in developing lung. J. Biol. Chem. 271: 11761-11766.
4. Maichele, A.J., et al. 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.
5. Burwinkel, B., et al. 1998. Liver glycogenosis due to phosphorylase kinase deficiency: PHKG2 gene structure and mutations associated with cirrhosis. Hum. Mol. Genet. 7: 149-154.
6. Burwinkel, B., et al. 2000. Phosphorylase kinase deficient liver glycogenosis: progression to cirrhosis in infancy associated with PHKG2 mutations (H144Y and L225R). J. Med. Genet. 37: 376-377.
7. Burwinkel, B., et al. 2003. Severe phenotype of phosphorylase kinase-deficient liver glycogenosis with mutations in the PHKG2 gene. Pediatr. Res. 54: 834-839.
8. 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.
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CHROMOSOMAL LOCATION

Genetic locus: PHKG1 (human) mapping to 7p11.2.

SOURCE

PHKG1 (RR-34) is a mouse monoclonal antibody raised against recombinant PHKG1 of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

PHKG1 (RR-34) is recommended for detection of PHKG1 of human origin by immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (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 PHKG1 siRNA (h): sc-89501, PHKG1 shRNA Plasmid (h): sc-89501-SH and PHKG1 shRNA (h) Lentiviral Particles: sc-89501-V.

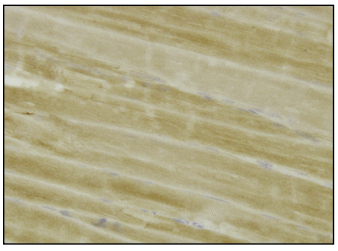
Molecular Weight of PHKG1: 45 kDa.

Positive Controls: Ramos cell lysate: sc-2216.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) 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. 2) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



PHKG1 (RR-34): sc-100536. Immunoperoxidase staining of formalin-fixed, paraffin-embedded human skeletal muscle tissue showing cytoplasmic localization.

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 for detailed protocols and support products.