SANTA CRUZ BIOTECHNOLOGY, INC.

PHKB (T-14): sc-160669

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Phosphorylase kinase is a hexadecameric enzyme that is comprised of four copies of four subunits that are encoded by four separate genes: PHKA, PHKB, PHKG and PHKD. This serine/threonine specific kinase converts glycogen phosphorylase β to glycogen phosphorylase α, resulting in the release of glucose-1-phophate from glycogen. PHKB (phosphorylase β kinase regulatory subunit β) is a 1093 amino acid subunit of phosphorylase kinase that, along with PHKA, has regulatory functions controlled by phosphorylation. Defects in the gene encoding PHKB are the cause of glycogen storage disease type 9B, which is also known as phosphorylase kinase deficiency of liver and muscle. This disease is characterized by a mild phenotype of hepatomegaly with only slightly elevated transaminase and plasma lipids, no clinical muscle involvement, and generally is correlated with a gradual improvement with increasing age. There are four isoforms of PHKB that are produced as a result of alternative splicing events.

REFERENCES

1. Wüllrich-Schmoll, A. and Kilimann, M.W. 1996. Structure of the human gene encoding the phosphorylase kinase β subunit (PHKB). Eur. J. Biochem. 238: 374-380.
2. van den Berg, I.E., et al. 1997. Autosomal recessive phosphorylase kinase deficiency in liver, caused by mutations in the gene encoding the β subunit (PHKB). Am. J. Hum. Genet. 61: 539-546.
3. Burwinkel, B., et al. 1997. Phosphorylase-kinase-deficient liver glycogenosis with an unusual biochemical phenotype in blood cells associated with a missense mutation in the β subunit gene (PHKB). Hum. Genet. 101: 170-174.
4. Burwinkel, B., et al. 2003. Muscle glycogenosis with low phosphorylase kinase activity: mutations in PHKA1, PHKG1 or six other candidate genes explain only a minority of cases. Eur. J. Hum. Genet. 11: 516-526.
5. Burwinkel, B., et al. 2003. Severe phenotype of phosphorylase kinase-deficient liver glycogenosis with mutations in the PHKG2 gene. Pediatr. Res. 54: 834-839.
6. Beauchamp, N.J., et al. 2007. Glycogen storage disease type IX: High variability in clinical phenotype. Mol. Genet. Metab. 92: 88-99.
7. Daub, H., et al. 2008. Kinase-selective enrichment enables quantitative phosphoproteomics of the kinome across the cell cycle. Mol. Cell 31: 438-448.
8. Online Mendelian Inheritance in Man, OMIM™. 2009. Johns Hopkins University, Baltimore, MD. MIM Number: 172490. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/

CHROMOSOMAL LOCATION

Genetic locus: PHKB (human) mapping to 16q12.1; Phkb (mouse) mapping to 8 C3.

SOURCE

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

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-160669 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

PHKB (T-14) is recommended for detection of PHKB 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).

PHKB (T-14) is also recommended for detection of PHKB in additional species, including equine, canine and bovine.

Suitable for use as control antibody for PHKB siRNA (h): sc-93503, PHKB siRNA (m): sc-152224, PHKB shRNA Plasmid (h): sc-93503-SH, PHKB shRNA Plasmid (m): sc-152224-SH, PHKB shRNA (h) Lentiviral Particles: sc-93503-V and PHKB shRNA (m) Lentiviral Particles: sc-152224-V.

Molecular Weight of PHKB: 125 kDa.

Positive Controls: Jurkat whole cell lysate: sc-2204.

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.

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.