

PHKA2 (F-1): sc-393491

BACKGROUND

PHKA2 (phosphorylase kinase, α 2), also known as PHKLA, XLG or PYK, is a 1,235 amino acid protein that is lipid-anchored to the cytoplasmic side of the cell membrane and belongs to the phosphorylase β kinase regulatory chain family. Expressed predominately in liver, but also present in other non-muscle tissues, PHKA2 exists as a component of a multi-chain polymer that functions as a phosphorylase β kinase and catalyzes the phosphorylation of target substrates, such as Troponin I. Defects in the gene encoding PHKA2 are the cause of glycogen storage disease type 9A (GSD9A), a metabolic disorder that results in glycogenosis (an abnormal accumulation of glycogen in tissue) and is characterized by hepatomegaly, growth retardation, muscle weakness, hypercholesterolemia, hypertriglyceridemia and fasting hyperketosis.

REFERENCES

1. Willems, P. 1990. Families with X-linked liver glycogenosis due to phosphorylase kinase deficiency. *Clin. Genet.* 38: 80.
2. Wauters, J.G., et al. 1992. Regional mapping of a liver α -subunit gene of phosphorylase kinase (PHKA) to the distal region of human chromosome Xp. *Cytogenet. Cell Genet.* 60: 194-196.
3. Hirono, H., et al. 1995. Isolation of cDNA encoding the human liver phosphorylase kinase α subunit (PHKA2) and identification of a missense mutation of the PHKA2 gene in a family with liver phosphorylase kinase deficiency. *Biochem. Mol. Biol. Int.* 36: 505-511.
4. Hendrickx, J., et al. 1995. Mutations in the phosphorylase kinase gene PHKA2 are responsible for X-linked liver glycogen storage disease. *Hum. Mol. Genet.* 4: 77-83.
5. Hendrickx, J., et al. 1996. X-linked liver glycogenosis type II (XLG II) is caused by mutations in PHKA2, the gene encoding the liver α subunit of phosphorylase kinase. *Hum. Mol. Genet.* 5: 649-652.

CHROMOSOMAL LOCATION

Genetic locus: PHKA2 (human) mapping to Xp22.13.

SOURCE

PHKA2 (F-1) is a mouse monoclonal antibody raised against amino acids 608-778 mapping within an internal region of PHKA2 of human origin.

PRODUCT

Each vial contains 200 μ g IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

PHKA2 (F-1) is available conjugated to agarose (sc-393491 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-393491 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-393491 PE), fluorescein (sc-393491 FITC), Alexa Fluor[®] 488 (sc-393491 AF488), Alexa Fluor[®] 546 (sc-393491 AF546), Alexa Fluor[®] 594 (sc-393491 AF594) or Alexa Fluor[®] 647 (sc-393491 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor[®] 680 (sc-393491 AF680) or Alexa Fluor[®] 790 (sc-393491 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

PHKA2 (F-1) is recommended for detection of PHKA2 of human origin by Western Blotting (starting dilution 1:100, 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).

Suitable for use as control antibody for PHKA2 siRNA (h): sc-76121, PHKA2 shRNA Plasmid (h): sc-76121-SH and PHKA2 shRNA (h) Lentiviral Particles: sc-76121-V.

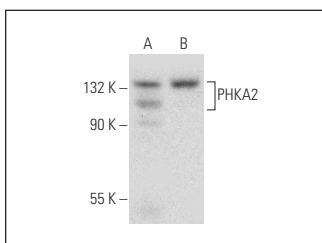
Molecular Weight of PHKA2: 138 kDa.

Positive Controls: human liver extract: sc-363766, Hep G2 cell lysate: sc-2227 or ZR-75-1 cell lysate: sc-2241.

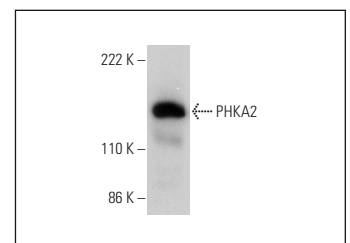
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker[™] Molecular Weight Standards: sc-2035, UltraCruz[®] Blocking Reagent: sc-516214 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 m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz[®] Mounting Medium: sc-24941 or UltraCruz[®] Hard-set Mounting Medium: sc-359850.

DATA



PHKA2 (F-1): sc-393491. Western blot analysis of PHKA2 expression in Hep G2 (A) and ZR-75-1 (B) whole cell lysates.



PHKA2 (F-1): sc-393491. Western blot analysis of PHKA2 expression in human liver tissue extract.

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.