

PHYH (E-19): sc-82021

BACKGROUND

PHYH (phytanoyl-CoA 2-hydroxylase), also known as RD, LN1, PAHX or LNP1, is a 338 amino acid protein that localizes to the peroxisome and plays an important role in fatty acid metabolism. Expressed in kidney, liver and T cells, PHYH uses iron and ascorbate as cofactors to catalyze the conversion of phytanoyl-CoA to 2-hydroxyphytanoyl-CoA, a reaction that is involved in the α -oxidation of 3-methyl branched fatty acids. Defects in the gene encoding PHYH are associated with Refsum disease (RD), an autosomal recessive disorder that is characterized by retinitis pigmentosa, peripheral neuropathy, cerebellar ataxia, nerve deafness, anosmia, skeletal abnormalities, ichthyosis, cataracts and cardiac impairment, all of which usually develop during the second or third decade of life.

REFERENCES

- Jansen, G.A., et al. 1997. Phytanoyl-coenzyme A hydroxylase deficiency—the enzyme defect in Refsum's disease. *N. Engl. J. Med.* 337: 133-134.
- Mihalik, S.J., et al. 1997. Identification of PAHX, a Refsum disease gene. *Nat. Genet.* 17: 185-189.
- Jansen, G.A., et al. 1999. Phytanoyl-CoA hydroxylase deficiency. Enzymological and molecular basis of classical Refsum disease. *Adv. Exp. Med. Biol.* 466: 371-376.
- Mukherji, M., et al. 2001. Structure-function analysis of phytanoyl-CoA 2-hydroxylase mutations causing Refsum's disease. *Hum. Mol. Genet.* 10: 1971-1982.
- Kee, H.J., et al. 2003. A novel murine long-chain acyl-CoA synthetase expressed in brain participates in neuronal cell proliferation. *Biochem. Biophys. Res. Commun.* 305: 925-933.

CHROMOSOMAL LOCATION

Genetic locus: PHYH (human) mapping to 10p13; Phyh (mouse) mapping to 2 A1.

SOURCE

PHYH (E-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of PHYH 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.

Blocking peptide available for competition studies, sc-82021 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

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.

APPLICATIONS

PHYH (E-19) is recommended for detection of PHYH 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).

PHYH (E-19) is also recommended for detection of PHYH in additional species, including canine and bovine.

Suitable for use as control antibody for PHYH siRNA (h): sc-76127, PHYH siRNA (m): sc-76128, PHYH shRNA Plasmid (h): sc-76127-SH, PHYH shRNA Plasmid (m): sc-76128-SH, PHYH shRNA (h) Lentiviral Particles: sc-76127-V and PHYH shRNA (m) Lentiviral Particles: sc-76128-V.

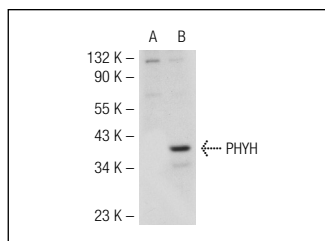
Molecular Weight of PHYH: 36 kDa.

Positive Controls: PHYH (m): 293T Lysate: sc-127330.

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

DATA



PHYH (E-19): sc-82021. Western blot analysis of PHYH expression in non-transfected: sc-117752 (A) and mouse PHYH transfected: sc-127330 (B) 293T whole cell lysates.

SELECT PRODUCT CITATIONS

- Kobayashi, Y., et al. 2010. Ameliorative effects of mulberry (*Morus alba* L.) leaves on hyperlipidemia in rats fed a high-fat diet: induction of fatty acid oxidation, inhibition of lipogenesis, and suppression of oxidative stress. *Biosci. Biotechnol. Biochem.* 74: 2385-2395.

RESEARCH USE

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