

PHYH (E-8): sc-376727

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

1. 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.
2. Mihalik, S.J., et al. 1997. Identification of PAHX, a Refsum disease gene. *Nat. Genet.* 17: 185-189.
3. 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.
4. Mukherji, M., et al. 2001. Structure-function analysis of phytanoyl-CoA 2-hydroxylase mutations causing Refsum's disease. *Hum. Mol. Genet.* 10: 1971-1982.

CHROMOSOMAL LOCATION

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

SOURCE

PHYH (E-8) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 145-181 within an internal region of PHYH 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.

PHYH (E-8) is available conjugated to agarose (sc-376727 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-376727 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-376727 PE), fluorescein (sc-376727 FITC), Alexa Fluor® 488 (sc-376727 AF488), Alexa Fluor® 546 (sc-376727 AF546), Alexa Fluor® 594 (sc-376727 AF594) or Alexa Fluor® 647 (sc-376727 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-376727 AF680) or Alexa Fluor® 790 (sc-376727 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-376727 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

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APPLICATIONS

PHYH (E-8) is recommended for detection of PHYH of mouse, rat and 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).

PHYH (E-8) is also recommended for detection of PHYH in additional species, including equine, 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.

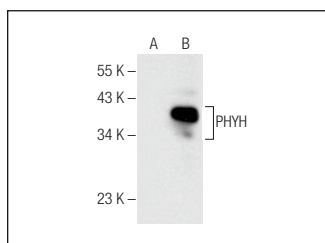
Molecular Weight of PHYH: 36 kDa.

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

RECOMMENDED 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



PHYH (E-8): sc-376727. Western blot analysis of PHYH expression in non-transfected: sc-117752 (A) and mouse PHYH transfected: sc-127330 (B) 293T whole cell lysates.

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.