



## PHYH (Y-14): sc-82023

### 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  $\alpha$ -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

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- Jansen, G.A., et al. 2004. Molecular basis of Refsum disease: sequence variations in phytanoyl-CoA hydroxylase (PHYH) and the PTS2 receptor (PEX7). *Hum. Mutat.* 23: 209-218.
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### CHROMOSOMAL LOCATION

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

### STORAGE

Store at 4° C, **\*\*DO NOT FREEZE\*\***. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

### SOURCE

PHYH (Y-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of PHYH of human origin.

### PRODUCT

Each vial contains 200  $\mu$ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-82023 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

### APPLICATIONS

PHYH (Y-14) 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), 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 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.

### 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.

### RESEARCH USE

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

### PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.