

# PAH (C-19): sc-15112

## BACKGROUND

The PAH gene encodes the enzyme phenylalanine hydroxylase (PAH), which converts phenylalanine to tyrosine and is the rate-limiting enzyme in phenylalanine catabolism. Mammalian PAH is a soluble, homotetrameric protein, which is abundantly expressed in human liver. Deficiency of PAH activity results in the autosomal recessive disorder phenylketonuria (PKU), which is characterized by mental retardation unless a low phenylalanine diet is introduced early in life. The PAH gene, which maps to human chromosome 12q23.2, contains all the genetic information necessary to code for functional PAH, demonstrating that a single gene is involved in the classic disease phenotype. Numerous mutations can impair the PAH gene, which result in decreased enzyme activity and give rise to varying degrees of PKU. Multiple isozymes of PAH have been reported to exist, but these are most likely allelic variants of PAH that produce protein subunits with slightly different charge and electrophoretic migration.

## CHROMOSOMAL LOCATION

Genetic locus: PAH (human) mapping to 12q23.2; Pah (mouse) mapping to 10 C1.

## SOURCE

PAH (C-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of PAH 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-15112 P, (100 µg 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.

## APPLICATIONS

PAH (C-19) is recommended for detection of PAH of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg 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).

PAH (C-19) is also recommended for detection of PAH in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for PAH siRNA (h): sc-41528, PAH siRNA (m): sc-41529, PAH shRNA Plasmid (h): sc-41528-SH, PAH shRNA Plasmid (m): sc-41529-SH, PAH shRNA (h) Lentiviral Particles: sc-41528-V and PAH shRNA (m) Lentiviral Particles: sc-41529-V.

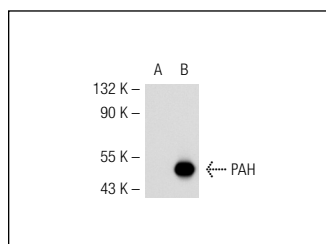
Molecular Weight of PAH: 51 kDa.

Positive Controls: PAH (m): 293T Lysate: sc-122353.

## 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



PAH (C-19): sc-15112. Western blot analysis of PAH expression in non-transfected: sc-117752 (A) and mouse PAH transfected: sc-122353 (B) 293T whole cell lysates.

## SELECT PRODUCT CITATIONS

1. Pan, Y.H., et al. 2013. Adaptation of phenylalanine and tyrosine catabolic pathway to hibernation in bats. PLoS ONE 8: e62039.

## 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.



Try **PAH (H-2): sc-271258** or **PAH (E-8): sc-271257**, our highly recommended monoclonal alternatives to PAH (C-19).