

PAH (PH8): sc-58398

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.

REFERENCES

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CHROMOSOMAL LOCATION

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

SOURCE

PAH (PH8) is a mouse monoclonal antibody raised against liver phenylalanine hydroxylase of monkey origin.

PRODUCT

Each vial contains 100 µg IgG₁ in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

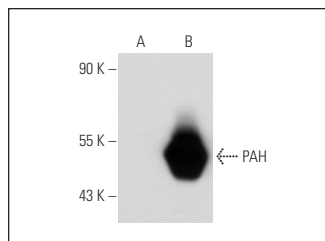
PAH (PH8) is recommended for detection of PAH of mouse, rat, human and monkey origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)] and immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500); also recommended for detection of serotonergic neurons in fixed tissues; is a marker for TYH-containing catecholaminergic neurons and serotonergic neurons in fresh tissue.

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.

DATA



PAH (PH8): sc-58398. Western blot analysis of PAH expression in non-transfected: sc-117752 (A) and mouse PAH transfected: sc-122353 (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.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.