SANTA CRUZ BIOTECHNOLOGY, INC.

PAH (H-2): sc-271258

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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

1. Hopkinson, D.A., et al. 1976. The distributions of subunit numbers and subunit sizes of enzymes: a study of the products of 100 human gene loci. Ann. Hum. Genet. 39: 383-411.
2. Ledley, F.D., et al. 1985. Gene transfer and expression of human phenylalanine hydroxylase. Science 228: 77-79.

CHROMOSOMAL LOCATION

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

SOURCE

PAH (H-2) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 404-432 near the C-terminus of PAH of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

PAH (H-2) is available conjugated to agarose (sc-271258 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-271258 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-271258 PE), fluorescein (sc-271258 FITC), Alexa Fluor® 488 (sc-271258 AF488), Alexa Fluor® 546 (sc-271258 AF546), Alexa Fluor® 594 (sc-271258 AF594) or Alexa Fluor® 647 (sc-271258 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-271258 AF680) or Alexa Fluor® 790 (sc-271258 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-271258 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

STORAGE

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

APPLICATIONS

PAH (H-2) is recommended for detection of PAH of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

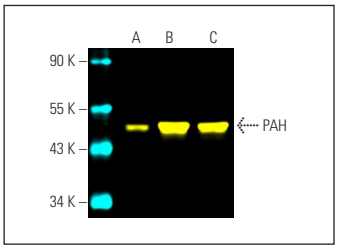
PAH (H-2) is also recommended for detection of PAH in additional species, including equine, canine and porcine.

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.

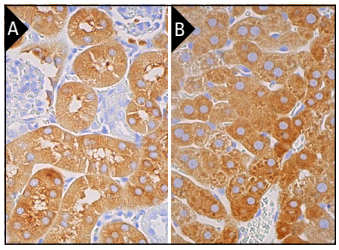
Molecular Weight of PAH: 51 kDa.

Positive Controls: human liver extract: sc-363766, mouse liver extract: sc-2256 or mouse kidney extract: sc-2255.

DATA



PAH (H-2) Alexa Fluor® 488: sc-271258 AF488. Direct fluorescent western blot analysis of PAH expression in human liver (**A**), mouse liver (**B**) and mouse kidney (**C**) tissue extracts. Blocked with UltraCruz® Blocking Reagent: sc-516214. Cruz Marker™ Molecular Weight Standards detected with Cruz Marker™ MW Tag-Alexa Fluor® 647: sc-516791.



PAH (H-2): sc-271258. Immunoperoxidase staining of formalin fixed, paraffin-embedded mouse kidney tissue showing cytoplasmic staining of cells in tubules (**A**), and of mouse liver tissue showing cytoplasmic staining of hepatocytes (**B**). Blocked with 0.25X UltraCruz® Blocking Reagent: sc-516214. Detection reagents used: m-IgGκ BP-B: sc-516142 and ImmunoCruz® ABC Kit: sc-516216.

SELECT PRODUCT CITATIONS

1. Ramírez, A.M., et al. 2017. Production of human recombinant phenylalanine hydroxylase in *Lactobacillus plantarum* for gastrointestinal delivery. Eur. J. Pharm. Sci. 109: 48-55.
2. Hendley, A.M., et al. 2021. Single-cell transcriptome analysis defines heterogeneity of the murine pancreatic ductal tree. Elife 10: e67776.
3. Vo, V.T.A., et al. 2022. Iron commensalism of mesenchymal glioblastoma promotes ferroptosis susceptibility upon dopamine treatment. Commun. Biol. 5: 593.
4. Yu, B.Y., et al. 2023. Dimethyl itaconate inhibits melanogenesis in B16F10 cells. Antioxidants 12: 692.

RESEARCH USE

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