PAH monoclonal antibody

Catalog: MB66750 Host: Mouse Reactivity: Human, Mouse, Rat

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.

Product:

Mouse IgG1 kappa. Liquid in PBS, pH 7.3, 30% glycerol, and 0.01% sodium azide.

Molecular Weight:

~ 52 kDa

Swiss-Prot:

P00439

Purification&Purity:

This antibody is purified through a protein G column.

Applications:

WB (1/500 - 1/1000), IHC (1/50 - 1/200)

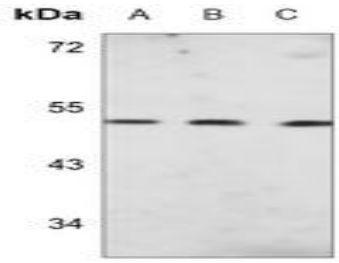
Storage&Stability:

Store at 4℃ short term. Aliquot and store at -20℃ long term. Avoid freeze-thaw cycles.

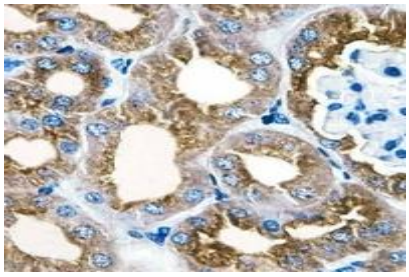
Specificity:

Recognizes endogenous levels of PAH protein.

DATA:



Western blot analysis of PAH expression in HepG2 (A), mouse liver (B), rat liver (C) whole cell lysates.



Immunohistochemical analysis of PAH staining in mouse kidney formalin fixed paraffin embedded tissue section. The section was pre-treated using heat mediated antigen retrieval with sodium citrate buffer (pH 6.0). The section was then incubated with the antibody at room temperature and detected using an HRP conjugated compact polymer system. DAB was used as the chromogen. The section was then counterstained with haematoxylin and mounted with DPX.

Note:

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

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