

PAH Rabbit Polyclonal Antibody

Catalog #: EAB14335

Host/Isotype	Clonality	Applications	MW (kDa)	Reactivity
Rabbit IgG	Polyclonal	WB, IHC-P, IF, ELISA	51	Human, Mouse, Rat

Applications Dilutions

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

WB (Western Blotting)	1:500-2000
IHC-P (Immunohistochemistry-Paraffin)	1:50-300
IF (Immunofluorescence)	1:50-300
ELISA (Enzyme-linked Immunosorbent Assay)	1:5000-20000

Product Information

Conjugate	Unconjugate
Specificity	PAH Rabbit Polyclonal Antibody detects endogenous levels of PAH protein.
Purification	Affinity purification
Concentration	1mg/ml
Format	Liquid
Formulation	In PBS, pH 7.4, Containing 0.02% sodium azide, 0.5% BSA and 50% Glycerol.
Shipping	Gel Pack
Storage	Store at -20°C least 1 year from the date of shipment. Avoid repeated freeze/thaw cycles. Aliquots may be stored at +4°C for 1-2 weeks.
UniProt ID	P00439
Entrez-Gene ID	5053

Product Description

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.

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