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Product datasheet for AP20792PU-M

PAH Rabbit Polyclonal Antibody

Product data:

Product Type:	Primary Antibodies
Applications:	IHC, WB
Recommended Dilution:	**Western blot:** 1/500 - 1/1000. **Immunohistochemistry on paraffin sections:** 1/50 - 1/200.
Reactivity:	Human, Mouse, Rat
Host:	Rabbit
Clonality:	Polyclonal
Specificity:	This antibody detects endogenous levels of PAH protein. (region surrounding Arg400)
Formulation:	Phosphate buffered saline (PBS), pH 7.2. State: Aff - Purified State: Liquid purified Ig fraction Preservative: 0.05% sodium azide
Concentration:	1.0 mg/ml
Purification:	Affinity chromatography (> 95% (by SDS-PAGE)
Conjugation:	Unconjugated
Storage:	Store undiluted at 2-8°C for one month or (in aliquots) at -20°C for longer. Avoid repeated freezing and thawing.
Stability:	Shelf life: one year from despatch.
Predicted Protein Size:	~ 55 kDa
Gene Name:	phenylalanine hydroxylase
Database Link:	Entrez Gene 18478 MouseEntrez Gene 24616 RatEntrez Gene 5053 Human P00439

View online »

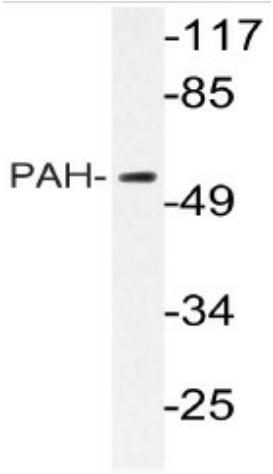
Background: The PAH gene encodes the enzyme phenylalanine hydroxylase (PAH), which converts phenylalanine to tyrosine and is the rate-limiting enzyme in phenylalanine catabolism. Mammalian PAH is a soluble, homotetrameric protein which is abundantly expressed in human liver. Deficiency of PAH activity 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.

Synonyms: Phenylalanine Hydroxylase, PH, PKU1, PKU

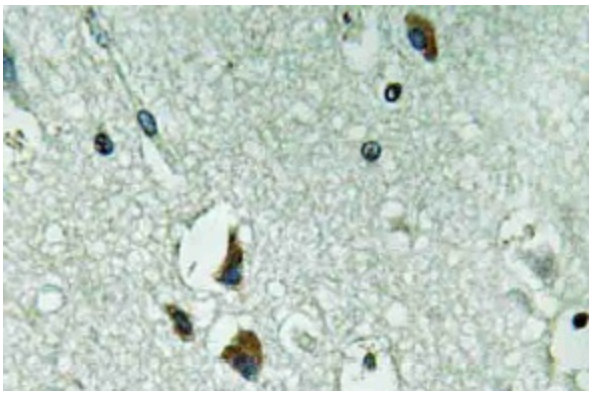
Protein Families: Druggable Genome

Protein Pathways: Metabolic pathways, Phenylalanine, tyrosine and tryptophan biosynthesis, Phenylalanine metabolism

Product images:



Western blot (WB) analyzes of PAH antibody (Cat.-No.: [AP20792PU-N]) in extracts from HepG2 cells.



Immunohistochemistry (IHC) analyzes of PAH antibody (Cat.-No.: [AP20792PU-N]) in paraffin-embedded human brain tissue.