abcam

Product datasheet

Anti-PAH antibody ab106805

2 Images

Overview

Product name	Anti-PAH antibody
Description	Goat polyclonal to PAH
Host species	Goat
Tested applications	**Suitable for:** WB
Species reactivity	**Reacts with:** Rat, Human **Predicted to work with:** Mouse, Chicken, Cow, Dog
Immunogen	Synthetic peptide: ESRPSRLKKDE by a Cysteine residue linker, corresponding to internal sequence amino acids 66-76 of Human PAH (NP_000268.1). Run BLAST with Run BLAST with
Positive control	WB: Human liver and rat kidney tissue lysates.
General notes	The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing. If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid repeated freeze / thaw cycles.
Storage buffer	pH: 7.30 Preservative: 0.02% Sodium azide Constituents: 99% Tris buffered saline, 0.5% BSA
Purity	Immunogen affinity purified
Purification notes	ab106805 is purified from Goat serum by ammonium sulphate precipitation followed by antigen affinity chromatography using the immunizing peptide.
Clonality	Polyclonal

Isotype IgG

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab106805 in the following tested applications.

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

Application	Abreviews	Notes
WB		Use a concentration of 0.1 - 0.3 µg/ml. Detects a band of approximately 48 kDa (predicted molecular weight: 52 kDa). 1 hour primary incubation is recommended for this product.

Target

Pathway Amino-acid degradation; L-phenylalanine degradation; acetoacetate and fumarate from L-phenylalanine: step 1/6.

Involvement in disease Defects in PAH are the cause of phenylketonuria (PKU) [MIM:261600]. PKU is an autosomal recessive inborn error of phenylalanine metabolism, due to severe phenylalanine hydroxylase deficiency. It is characterized by blood concentrations of phenylalanine persistently above 1200 mumol (normal concentration 100 mumol) which usually causes mental retardation (unless low phenylalanine diet is introduced early in life). They tend to have light pigmentation, rashes similar to eczema, epilepsy, extreme hyperactivity, psychotic states and an unpleasant 'mousy' odor.
Defects in PAH are the cause of non-phenylketonuria hyperphenylalaninemia (Non-PKU HPA) [MIM:261600]. Non-PKU HPA is a mild form of phenylalanine hydroxylase deficiency characterized by phenylalanine levels persistently below 600 mumol, which allows normal intellectual and behavioral development without treatment. Non-PKU HPA is usually caused by the combined effect of a mild hyperphenylalaninemia mutation and a severe one.
Defects in PAH are the cause of hyperphenylalaninemia (HPA) [MIM:261600]. HPA is the mildest form of phenylalanine hydroxylase deficiency.

Sequence similarities Belongs to the biopterin-dependent aromatic amino acid hydroxylase family.
Contains 1 ACT domain.

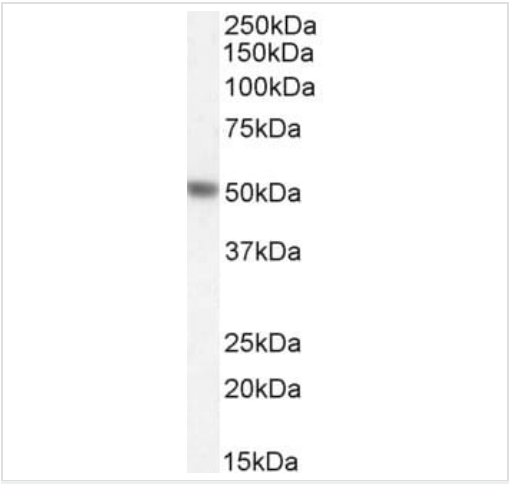
Images

Western blot - Anti-PAH antibody (ab106805)

Anti-PAH antibody (ab106805) at 0.1 µg/ml + Human liver tissue lysate (35µg protein in RIPA buffer)

Developed using the ECL technique.

Predicted band size: 52 kDa



Western blot - Anti-PAH antibody (ab106805)

Anti-PAH antibody (ab106805) at 0.3 µg/ml + Rat kidney tissue lysate (35µg protein in RIPA buffer)

Developed using the ECL technique.

Predicted band size: 52 kDa

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery

- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit **https://www.abcam.com/abpromise** or contact our technical team.

Terms and conditions

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors