abcam

Product datasheet

Anti-PAH antibody ab106805

2 图像

概述

产品名称	Anti-PAH抗体
描述	**山羊多克隆抗体**to PAH
宿主	Goat
经测试应用	**适用于:** WB
种属反应性	**与反应:** Rat, Human **预测可用于:** Mouse, Chicken, Cow, Dog
免疫原	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
阳性对照	WB: Human liver and rat kidney tissue lysates.
常规说明	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

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid repeated freeze / thaw cycles.
存储溶液	pH: 7.30 Preservative: 0.02% Sodium azide Constituents: 99% Tris buffered saline, 0.5% BSA
纯度	Immunogen affinity purified
纯化说明	ab106805 is purified from Goat serum by ammonium sulphate precipitation followed by antigen affinity chromatography using the immunizing peptide.
克隆	**多克隆**

同种型 IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用**ab106805**于以下的**经测试应用

“应用说明”部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
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.

靶标

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

疾病相关 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.

序列相似性 Belongs to the biopterin-dependent aromatic amino acid hydroxylase family.
Contains 1 ACT domain.

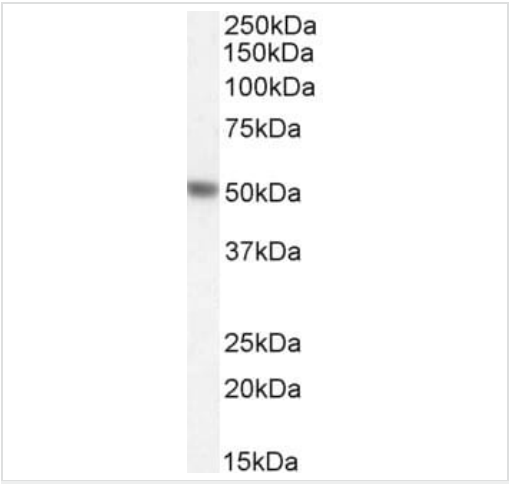
图片

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

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