


Anti-PAH antibody ab106805

画像数 2

製品の概要

製品名	Anti-PAH antibody
製品の詳細	Goat polyclonal to PAH
由来種	Goat
アプリケーション	適用あり: WB
種交差性	交差種: Rat, Human 交差が予測される動物種: 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
ポジティブ・コントロール	WB: Human liver and rat kidney tissue lysates.
特記事項	<p>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.</p> <p>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</p>

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid repeated freeze / thaw cycles.
バッファー	pH: 7.30 Preservative: 0.02% Sodium azide Constituents: 99% Tris buffered saline, 0.5% BSA
精製度	Immunogen affinity purified
特記事項(精製)	ab106805 is purified from Goat serum by ammonium sulphate precipitation followed by antigen affinity chromatography using the immunizing peptide.
ポリ/モノ	ポリクローナル

アプリケーション

The Abpromise guarantee **Abpromise保証は、** 次のテスト済みアプリケーションにおけるab106805の使用に適用されます
アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
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.

ターゲット情報

パスウェイ

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

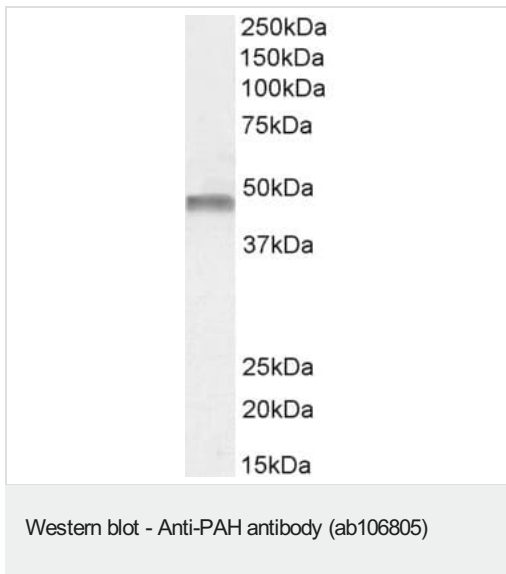
関連疾患

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 µmol (normal concentration 100 µmol) 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 µmol, 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.

配列類似性

Belongs to the bipterin-dependent aromatic amino acid hydroxylase family.
Contains 1 ACT domain.

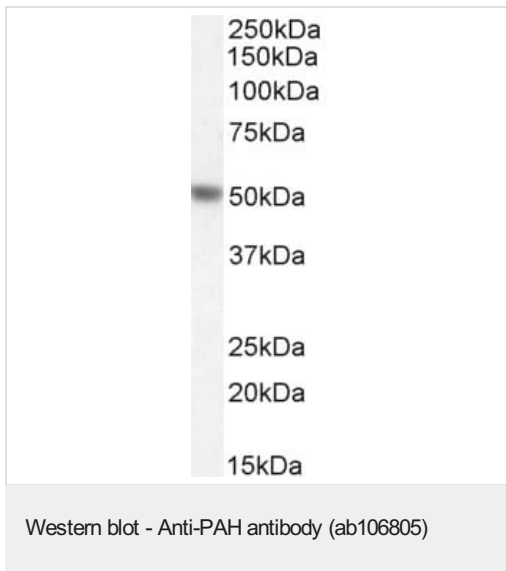
画像



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



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

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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