

Anti-PAH antibody ab88740

3 图像

概述

产品名称	Anti-PAH抗体
描述	小鼠多克隆抗体to PAH
宿主	Mouse
经测试应用	适用于: WB, IHC-P
种属反应性	与反应: Human
免疫原	Recombinant full length protein within Human PAH. The exact immunogen sequence used to generate this antibody is proprietary information. If additional detail on the immunogen is needed to determine the suitability of the antibody for your needs, please contact our Scientific Support team to discuss your requirements.
阳性对照	WB: Human liver tissue lysate; PAH transfected 293T cell lysate IHC-P: Human kidney tissue
常规说明	<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>

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
存储溶液	pH: 7.40 Constituent: 100% PBS
纯度	Protein A purified
克隆	多克隆
同种型	IgG

应用

The Abpromise guarantee

Abpromise™ 承诺保证使用ab88740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		
IHC-P		

应用说明

IHC-P: Use at a concentration of 3 µg/ml. Antigen retrieval is recommended. Place sample in 1X citrate buffer (pH 6.0) and microwave at 750W for 20 minutes, cool sample
WB: Use at a concentration of 1 µg/ml. Detects a band of approximately 50 kDa (predicted molecular weight: 52 kDa).

Not yet tested in other applications.

Optimal dilutions/concentrations should be determined by the end user.

靶标

通路

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

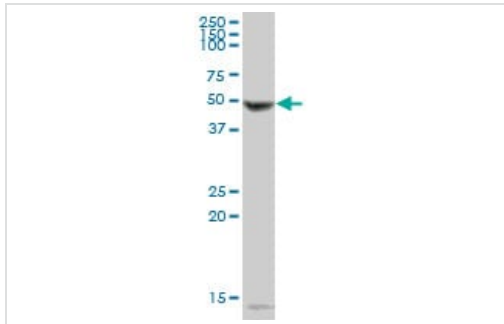
疾病相关

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 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.

序列相似性

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

图片



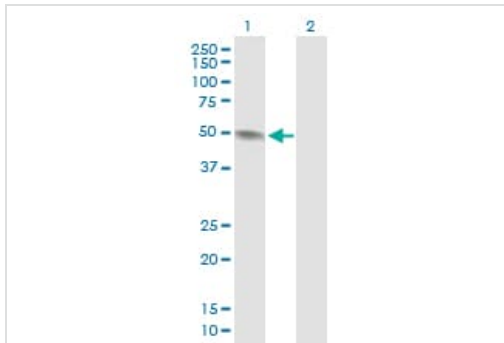
Western blot - Anti-PAH antibody (ab88740)

Anti-PAH antibody (ab88740) at 1 µg/ml + Human liver tissue lysate at 50 µg

Predicted band size: 52 kDa

Observed band size: 50 kDa

Additional bands at: <15 kDa. We are unsure as to the identity of these extra bands.



Western blot - Anti-PAH antibody (ab88740)

All lanes : Anti-PAH antibody (ab88740) at 1 µg/ml

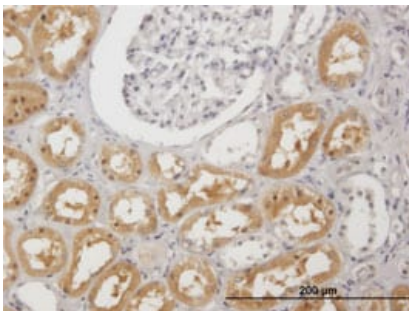
Lane 1 : PAH transfected 293T cell lysate

Lane 2 : Non transfected 293T cell lysate

Lysates/proteins at 25 µg per lane.

Predicted band size: 52 kDa

Observed band size: 50 kDa



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-PAH antibody (ab88740)

ab88740, at 3 µg/ml, staining PAH in formalin fixed, paraffin embedded Human kidney tissue by Immunoperoxidase/ Immunohistochemistry.

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

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