abcam

Product datasheet

Anti-Fumarylacetoacetate hydrolase/FAA antibody ab81087

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概述

产品名称	Anti-Fumarylacetoacetate hydrolase/FAA**抗体**
描述	**兔多克隆抗体**to Fumarylacetoacetate hydrolase/FAA
宿主	Rabbit
经测试应用	**适用于:** ICC/IF, WB, IP, ELISA, IHC-P
种属反应性	**与反应:** Mouse **预测可用于:** Rat, Human
免疫原	A full length recombinant protein of Fumarylacetoacetate hydrolase from Mouse origin.
阳性对照	Mouse liver tissue. Whole cell lysate derived from mouse liver tissue.

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	Preservative: None Constituents: PBS, pH 7.2, containing antibody stabilizer.
纯度	Immunogen affinity purified
克隆	**多克隆**
同种型	IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用**ab81087**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
ICC/IF	(1)	Use a concentration of 5 µg/ml.
WB		Use a concentration of 0.1 - 1 µg/ml. Detects a band of approximately 47 kDa (predicted molecular weight: 46 kDa).

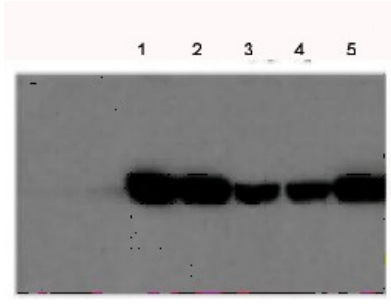
应用	Ab评论	说明
IP		Use a concentration of 2 - 5 µg/ml.
ELISA		Use a concentration of 0.01 - 0.1 µg/ml.
IHC-P		Use a concentration of 2 - 5 µg/ml.

靶标

相关性

Fumarylacetoacetate hydrolase / FAA is the last enzyme in the tyrosine catabolism pathway. FAH deficiency is associated with Type 1 hereditary tyrosinemia (HT). This is an autosomal recessive inborn error of metabolism that occurs in both an acute and a chronic form. Clinical characteristics of the acute form include hepatic failure and death in infancy, whereas children with the chronic form have renal tubular dysfunction and hypophosphatemic rickets, progressive liver disease with development of hepatocellular carcinoma. Dietary treatment with restriction of tyrosine and phenylalanine alleviates the rickets, but liver transplantation has so far been the only definite treatment.

图片

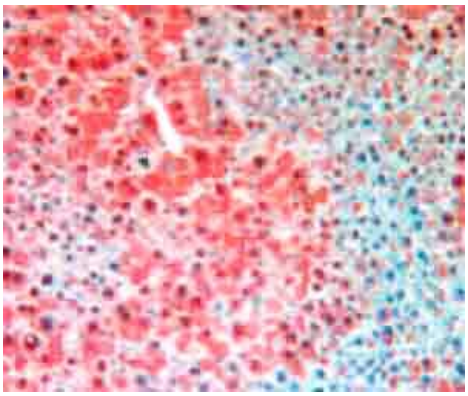


Western blot - Anti-Fumarylacetoacetate hydrolase antibody (ab81087)

All lanes : Anti-Fumarylacetoacetate hydrolase/FAA antibody (ab81087) at 1/500 dilution

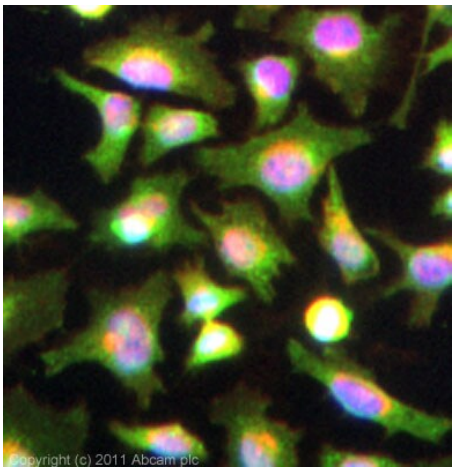
Lane 1 : Whole cell lysate derived from liver extract from FAH wild type mice
Lane 2 : Whole cell lysate derived from liver extract from FAH wild type mice
Lane 3 : Whole cell lysate derived from FAH-liver extract from z alleles transgenic mice
Lane 4 : Whole cell lysate derived from FAH-liver extract from z alleles transgenic mice
Lane 5 : Whole cell lysate derived from liver extract from siLucFAH mice

Predicted band size: 46 kDa
Observed band size: 47 kDa



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Fumarylacetoacetate hydrolase antibody (ab81087)

ab81087 at 2µg/ml staining Fumarylacetoacetate hydrolase by Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded mouse liver sections), followed by HRP linked secondary antibody, and visualized by AEC substrate, and counterstaining by hematoxylin.



Immunocytochemistry/ Immunofluorescence - Anti-Fumarylacetoacetate hydrolase antibody (ab81087)

ICC/IF image of ab81087 stained HeLa cells. The cells were 4% PFA fixed (10mins) and then incubated in 1%BSA / 10% normal goat serum / 0.3M glycine in 0.1% PBS-Tween for 1h to permeabilise the cells and block non-specific protein-protein interactions. The cells were then incubated with the antibody (ab81087, 5µg/ml) overnight at +4°C. The secondary antibody (green) was **ab96899** Dylight 488 goat anti-rabbit IgG (H+L) used at a 1/250 dilution for 1h. Alexa Fluor® 594 WGA was used to label plasma membranes (red) at a 1/200 dilution for 1h. DAPI was used to stain the cell nuclei (blue) at a concentration of 1.43µM.

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