

Product datasheet

Anti-Fumarylacetoacetate hydrolase antibody [2] ab83078

1 图像

概述

产品名称	Anti-Fumarylacetoacetate hydrolase 抗体[2]
描述	小鼠单克隆抗体[2] to Fumarylacetoacetate hydrolase
宿主	Mouse
经测试应用	适用于: WB, ELISA
种属反应性	与反应: Human
免疫原	Recombinant full length Human Fumarylacetoacetate hydrolase protein expressed in <i>E. coli</i> .
阳性对照	HEK293 whole cell lysate

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	Preservative: 0.02% Proclin Constituents: Tris HCl, Sodium citrate, pH 7.0
纯度	Protein A purified
克隆	单克隆
克隆编号	2
同种型	IgG2b

应用

Our [Abpromise guarantee](#) covers the use of **ab83078** in the following tested applications.

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

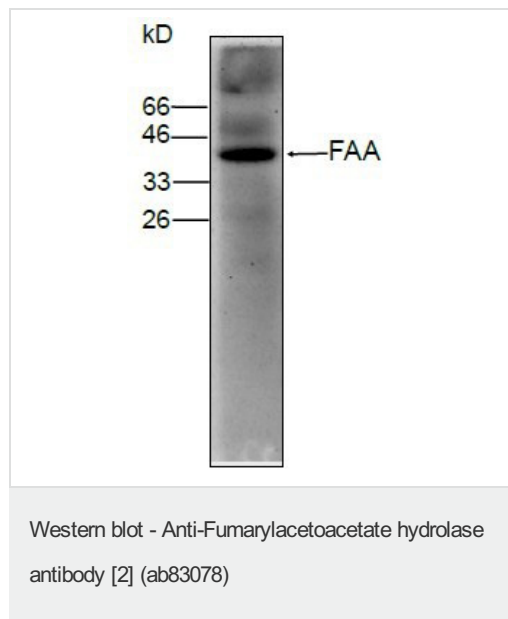
应用	Ab评论	说明
WB		1/1000. Predicted molecular weight: 46 kDa.
ELISA		Use at an assay dependent dilution.

## 靶标

### 相关性

Fumarylacetoacetate hydrolase 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



Anti-Fumarylacetoacetate hydrolase antibody [2] (ab83078) at 1/500 dilution + Whole cell lysates prepared from HEK293 cells.

**Predicted band size: 46 kDa**

**Please note:** All products are "FOR RESEARCH USE ONLY AND ARE NOT INTENDED FOR DIAGNOSTIC OR THERAPEUTIC USE"

### 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.cn/abpromise> or contact our technical team.

### Terms and conditions

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