

## Product datasheet

# Anti-Fumarylacetoacetate hydrolase antibody [2] ab83078

### 画像数 1

#### 製品の概要

製品名	Anti-Fumarylacetoacetate hydrolase antibody [2]
製品の詳細	Mouse monoclonal [2] to Fumarylacetoacetate hydrolase
由来種	Mouse
アプリケーション	適用あり: WB, ELISA
種交差性	交差種: Human
免疫原	Recombinant full length Human Fumarylacetoacetate hydrolase protein expressed in <i>E. coli</i> .
ポジティブ・コントロール	HEK293 whole cell lysate

#### 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	Preservative: 0.02% Proclin Constituents: Tris HCl, Sodium citrate, pH 7.0
精製度	Protein A purified
ポリ/モノ	モノクローナル
クローン名	2
アイソタイプ	IgG2b

#### アプリケーション

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.

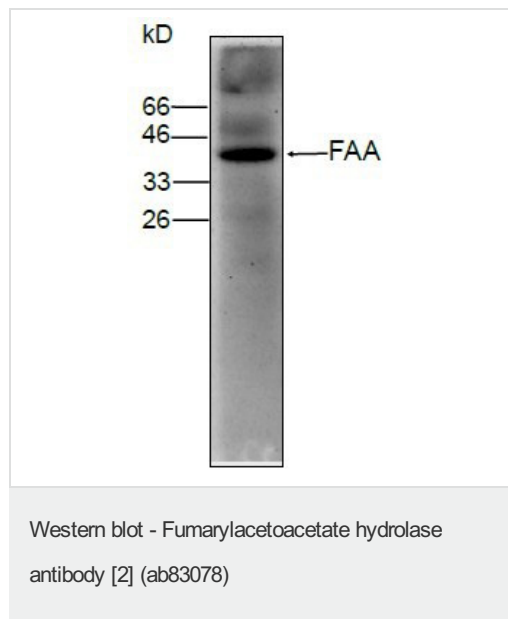
アプリケーション	Abreviews	特記事項
WB		1/1000. Predicted molecular weight: 46 kDa.
ELISA		Use at an assay dependent dilution.

## ターゲット情報

### 関連性

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.

### 画像



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"

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