

# Anti-HIBADH 兔多克隆抗体

K1335136 -20°C



## 产品信息

蛋白质全称	3-羟基异丁酸脱氢酶
别名	HIBADH, mitochondrial, NS5ATP1, EC 1.1.1.31, MGC40361
免疫原	小鼠 HIBADH 重组蛋白
Uniprot ID	Q99L13
抗体亚型	IgG
纯化方式	亲和纯化
预测分子量 / 观测分子量	35 kDa / 30 kDa

## 应用

### WB 免疫印迹

物种：小鼠，大鼠

稀释比：1: 500-1: 1000

阳性样品：肾

## 背景

HIBADH is a 336 amino acid mitochondrial enzyme that catalyzes the NAD<sup>+</sup>-dependent, reversible oxidation of 3-Hydroxyisobutyrate to methylmalonate semialdehyde, an intermediate of valine catabolism. The enzyme functions as a homodimer between a pH of 7.0 and 10.0, with optimal activity between 8.8 and 9.0. It was previously hypothesized that defects in the gene encoding HIBADH may be the cause of 3-Hydroxyisobutyric aciduria, a rare disorder that is characterized by a variety of clinical manifestations such as neurodevelopmental problems and dysmorphic features.

## 储存条件

**储存条件：**自收到之日起 -20°C下储存一年，避免反复冻融。  
**储存缓冲液：**含 0.15% ProClin300 防腐剂, 100 μg/mL 牛血清白蛋白和 50%甘油的磷酸盐缓冲液。

## 图像



WB 检测 HIBADH 蛋白(货号 K1335136).  
 样品: 经 RIPA 裂解液(货号 KSG2002)处理的蛋白质.  
 封闭: 3%脱脂牛奶(货号 KSGC310001)溶于 TBST 溶液,  
 室温孵育 1 小时.  
 一抗: 1: 1000 稀释, 4°C 孵育过夜.  
 二抗: HRP 标记山羊抗兔 IgG (H+L) (货号 KB69909), 1:  
 5000 稀释, 室温孵育 1 小时.

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