abcam

Product datasheet

Anti-ACADVL/VLCAD antibody ab99490

1 图像

概述

产品名称	Anti-ACADVL/VLCAD抗体
描述	**山羊多克隆抗体**to ACADVL/VLCAD
宿主	Goat
特异性	This antibody is expected to recognize both reported isoforms (NP_000009.1; NP_001029031.1).
经测试应用	**适用于:** WB
种属反应性	**与反应:** Human
免疫原	Synthetic peptide corresponding to N terminal amino acids 50-63 (DKSDSHPSDALTRK) of Human ACADVL (NP_000009.1). Run BLAST with Run BLAST with
阳性对照	Human heart 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% Sodium Azide Constituents: 0.5% BSA, Tris buffered saline, pH 7.3
纯度	Immunogen affinity purified
纯化说明	Purified from goat serum by ammonium sulphate precipitation followed by antigen affinity chromatography using the immunizing peptide.
克隆	**多克隆**
同种型	IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用**ab99490**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		Use a concentration of 1 - 3 µg/ml. Detects a band of approximately 68 kDa (predicted molecular weight: 68 kDa).

靶标

功能	Active toward esters of long-chain and very long chain fatty acids such as palmitoyl-CoA, mysritoyl-CoA and stearoyl-CoA. Can accomodate substrate acyl chain lengths as long as 24 carbons, but shows little activity for substrates of less than 12 carbons.
通路	Lipid metabolism; mitochondrial fatty acid beta-oxidation.
疾病相关	Defects in ACADVL are the cause of acyl-CoA dehydrogenase very long chain deficiency (ACADVLD) [MIM:201475]. ACADVLD is an autosomal recessive disease which leads to impaired long-chain fatty acid beta-oxidation. It is clinically heterogeneous, with three major phenotypes: a severe childhood form, with early onset, high mortality, and high incidence of cardiomyopathy; a milder childhood form, with later onset, usually with hypoketotic hypoglycemia as the main presenting feature, low mortality, and rare cardiomyopathy; and an adult form, with isolated skeletal muscle involvement, rhabdomyolysis, and myoglobinuria, usually triggered by exercise or fasting.
序列相似性	Belongs to the acyl-CoA dehydrogenase family.
细胞定位	Mitochondrion inner membrane.

图片

Western blot - Anti-ACADVL antibody (ab99490)

Anti-ACADVL/VLCAD antibody (ab99490) at 2 µg/ml + Human heart lysate (in RIPA buffer) at 35 µg

Predicted band size: 68 kDa

Incubation with ab99490 was 1 hour.

Detected by chemiluminescence.

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