# abcam

## Product datasheet

## Anti-ACADVL/VLCAD antibody [6A9AF2] ab110285

1 References 3 图像

概述

产品名称 Anti-ACADVL/VLCAD抗体[6A9AF2]

**小**鼠单**克隆抗体**[6A9AF2] to ACADVL/VLCAD

**宿主** Mouse

经测试应用 适用于: ICC/IF, IP, IHC-P

种属反应性 与反应: Human

免疫原 Tissue, cells or virus. This information is considered to be commercially sensitive.

阳性对照 Human HDFn cells; Human liver mitochondria; Human cerebellum tissue.

常规说明

This antibody clone is manufactured by Abcam. If you require a custom buffer formulation or

conjugation for your experiments, please contact orders@abcam.com.

The Life Science industry has been in the grips of a reproducibility crisis for a number of years.

Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets

your needs before purchasing.

If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be

found below, along with publications, customer reviews and Q&As

Product was previously marketed under the MitoSciences sub-brand.

性能

形式 Liquid

**存放说明** Shipped at 4°C. Store at +4°C. Do Not Freeze.

**存储溶液** pH: 7.5

Preservative: 0.02% Sodium azide Constituent: HEPES buffered saline

纯**度** Proprietary Purification

纯**化说明** The antibody was produced in vitro using hybridoma grown in serum-free media and then purified

by biochemical fractionation. Purity >95% by SDS-PAGE.

**克隆** 单克隆

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**克隆编号** 6A9AF2

**同种型** lgG1

轻链类型 kappa

#### 应用

### The Abpromise guarantee Abpromise™承诺保证使用ab110285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		Use a concentration of 5 µg/ml.
IP		Use at an assay dependent concentration.
IHC-P		1/250. Perform heat mediated antigen retrieval - 1 min pressure cook in 1mmol EDTA pH8.

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

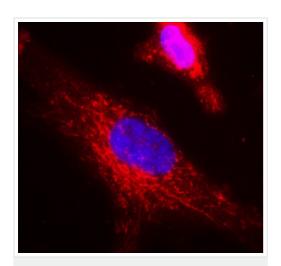
(ACADVLD) [MIM:201475]. ACADVLD is an autosomal recessive disease which leads to impaired long-chain fatty acid beta-oxidation. It is clinically heterogeneous, with three major phenotypes: a severe childhood form, with early onset, high mortality, and high incidence of cardiomyopathy; a milder childhood 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.

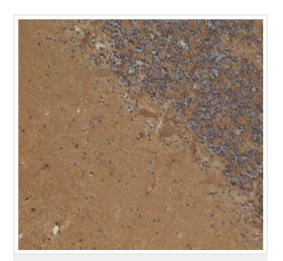
细**胞定位** Mitochondrion inner membrane.

图片



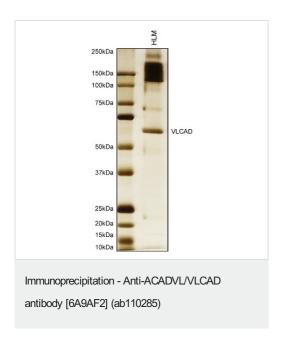
Immunocytochemistry/ Immunofluorescence - Anti-ACADVL/VLCAD antibody [6A9AF2] (ab110285)

Immunocytochemistry image of ACADVL/VLCAD antibody [6A9AF2] (ab110285) stained human HDFn cells. The cells were paraformaldehyde fixed (4%, 20 min) and Triton X-100 permeabilized (0.1%, 15min). The cells were incubated with ab110285 at 5 µg/ml for 2h at room temperature or over night at 4°C. The secondary antibody was (red) Alexa Fluor® 594 goat antimouse lgG (H+L) used at a 1/1000 dilution for 1h. 10% Goat serum was used as the blocking agent for all blocking steps. DAPI was used to stain the cell nuclei (blue). Target protein locates mainly in mitochondria.



Immunohistochemistry (Formalin/PFA-fixed paraffinembedded sections) - Anti-ACADVL/VLCAD antibody [6A9AF2] (ab110285)

ab110285,at 1/250 dilution, staining ACADVL/VLCAD in formalin-fixed, paraffin-embedded human cerebellum tissue. Immunoactivity is most intense in neuronal cell bodies, most notably in the large Purkinje cells.



ACADVL/VLCAD immunocaptured from 0.75mg of human liver mitochondria lysate /10ul antibody conjugated beads using ab110285.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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