abcam

Product datasheet

Recombinant Human ACADL/LCAD protein ab114591

1 图像

描述

产品名称 重组人ACADL/LCAD蛋白

表达系统 Wheat germ Accession P28330

蛋白长度 Full length protein

无动物成分 No

性质 Recombinant

种属 Human

序列 MAARLLRGSLRVLGGHRAPRQLPAARCSHSGGEERLETPSAK

KLTDIGIR

RIFSPEHDIFRKSVRKFFQEEVIPHHSEWEKAGEVSREVWEK

AGKQGLLG

VNIAEHLGGIGGDLYSAAIVWEEQAYSNCSGPGFSIHSGIVM

SYITNHGS

EEQIKHFIPQMTAGKCIGAIAMTEPGAGSDLQGIKTNAKKDG

SDWILNGS

KVFISNGSLSDVVIVVAVTNHEAPSPAHGISLFLVENGMKGF

IKGRKLHK

MGLKAQDTAELFFEDIRLPASALLGEENKGFYYIMKELPQER

LLIADVAI

 ${\tt SASEFMFEETRNYVKQRKAFGKTVAHLQTVQHKLAELKTHIC}$

VTRAFVDN

CLQLHEAKRLDSATACMAKYWASELQNSVAYDCVQLHGGWGY MWEYPIAK AYVDARVQPIYGGTNEIMKELIAREIVFDK

预**测分子量** 73 kDa including tags

氨基酸 1 to 430

技术指标

Our Abpromise quarantee covers the use of ab114591 in the following tested applications.

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

应**用** ELISA

SDS-PAGE

Western blot

形式 Liquid

补充说明 This product was previously labelled as ACADL.

制备和贮存

稳定性和存储 Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.

pH: 8.00

Constituents: 0.3% Glutathione, 0.79% Tris HCI

常规信息

通路 Lipid metabolism; mitochondrial fatty acid beta-oxidation.

疾病相关 Defects in ACADL are a cause of acyl-CoA dehydrogenase very long-chain deficiency

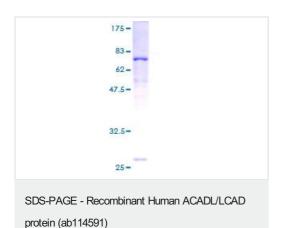
(ACADVLD) [MIM:201475]. An inborn error of mitochondrial fatty acid beta-oxidation which leads to impaired long-chain fatty acid beta-oxidation. It is clinically heterogeneous, with three major phenotypes: a severe childhood form characterized by early onset, high mortality and high incidence of cardiomyopathy; a milder childhood form with later onset, characterized by hypoketotic hypoglycemia, low mortality and rare cardiomyopathy; 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 matrix.

图片



12.5% SDS-PAGE showing ab114591 at approximately 73.41kDa stained with Coomassie Blue.

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