

ECHA rabbit pAb

Cat No.: ES16812

For research use only

Overview

Product Name ECHA rabbit pAb

Host species Rabbit Applications WB

Species Cross-Reactivity Human; Mouse;Rat Recommended dilutions WB 1:500-2000

Immunogen Synthesized peptide derived from human ECHA AA

range: 276-326

Specificity This antibody detects endogenous levels of ECHA at

Human/Mouse/Rat

Formulation Liquid in PBS containing 50% glycerol, 0.5% BSA and

0.02% sodium azide.

Storage Store at -20°C. Avoid repeated freeze-thaw cycles.

Protein Name ECHA

Gene Name HADHA HADH

Cellular localization Mitochondrion . Mitochondrion inner membrane .

Protein stability and association with mitochondrion

inner membrane do not require HADHB. .

Purification The antibody was affinity-purified from rabbit

antiserum by affinity-chromatography using

epitope-specific immunogen.

Clonality Polyclonal
Concentration 1 mg/ml
Observed band 85kD
Human Gene ID 3030
Human Swiss-Prot Number P40939

Alternative Names Trifunctional enzyme subunit alpha, mitochondrial

(78 kDa gastrin-binding protein) (TP-alpha)
[Includes: Long-chain enoyl-CoA hydratase (EC 4.2.1.17); Long chain 3-hydroxyacyl-CoA

dehydrogenase (EC 1.1.1.211)]

Background This gene encodes the alpha subunit of the

mitochondrial trifunctional protein, which catalyzes the last three steps of mitochondrial beta-oxidation

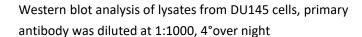


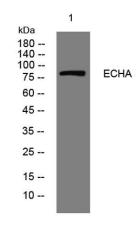
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of long chain fatty acids. The mitochondrial membrane-bound heterocomplex is composed of four alpha and four beta subunits, with the alpha subunit catalyzing the 3-hydroxyacyl-CoA dehydrogenase and enoyl-CoA hydratase activities. Mutations in this gene result in trifunctional protein deficiency or LCHAD deficiency. The genes of the alpha and beta subunits of the mitochondrial trifunctional protein are located adjacent to each other in the human genome in a head-to-head orientation. [provided by RefSeq, Jul 2008],







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