

Recombinant Human HADHA cell lysate

Cat. No. HADHA-2120HCL Lot. No. (See product label)

SPECIFICATION

Product Overview

Species	Human
Description	This gene encodes the alpha subunit of the mitochondrial trifunctional protein, which catalyzes the last three steps of mitochondrial beta-oxidation 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.
Size	100 ul
Storage Buffer	1X Sample Buffer (50 mM Tris-HCl, 2% SDS, 10% glycerol, 300 mM 2-mercaptoethanol, 0.01% Bromophenol blue)
Applications	Western Blot;

GENE INFORMATION

Gene Name	HADHA hydroxyacyl-CoA dehydrogenase/3-ketoacyl-CoA thiolase/enoyl-CoA hydratase (trifunctional protein), alpha subunit [Homo sapiens]
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Official Symbol	HADHA
Synonyms	HADHA; hydroxyacyl-CoA dehydrogenase/3-ketoacyl-CoA thiolase/enoyl-CoA hydratase (trifunctional protein), alpha subunit; hydroxyacyl Coenzyme A dehydrogenase/3 ketoacyl Coenzyme A thiolase/enoyl Coenzyme A hydratase (trifunctional protein), alpha subunit; trifunctional enzyme subunit alpha, mitochondrial; gastrin binding protein; GBP; LCEH; LCHAD; long chain 2 enoyl CoA hydratase; long chain 3 hydroxyacyl CoA dehydrogenase; mitochondrial trifunctional protein; alpha subunit; MTPA; 3-oxoacyl-CoA thiolase; gastrin-binding protein; 78 kDa gastrin-binding protein; long-chain 2-enoyl-CoA hydratase; long-chain-3-hydroxyacyl-CoA dehydrogenase; mitochondrial trifunctional enzyme, alpha subunit; mitochondrial trifunctional protein, alpha subunit; 3-ketoacyl-Coenzyme A (CoA) thiolase, alpha subunit; mitochondrial long-chain 2-enoyl-Coenzyme A (CoA) hydratase, alpha subunit; mitochondrial long-chain L-3-hydroxyacyl-Coenzyme A (CoA) dehydrogenase, alpha subunit; hydroxyacyl-Coenzyme A dehydrogenase/3-ketoacyl-Coenzyme A thiolase/enoyl-Coenzyme A hydratase (trifunctional protein), alpha subunit; ECHA; HADH; TP-ALPHA; MGC1728;
Gene ID	3030
mRNA Refseq	NM_000182
Protein Refseq	NP_000173
MIM	600890
UniProt ID	P40939
Chromosome Location	2p23

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Pathway

Beta oxidation of decanoyl-CoA to octanoyl-CoA-CoA, organism-specific biosystem;
 Beta oxidation of hexanoyl-CoA to butanoyl-CoA, organism-specific biosystem; Beta
 oxidation of lauroyl-CoA to decanoyl-CoA-CoA, organism-specific biosystem; Beta
 oxidation of myristoyl-CoA to lauroyl-CoA, organism-specific biosystem; Beta
 oxidation of octanoyl-CoA to hexanoyl-CoA, organism-specific biosystem; Beta
 oxidation of palmitoyl-CoA to myristoyl-CoA, organism-specific biosystem;
 Biosynthesis of unsaturated fatty acids, organism-specific biosystem;

Function

3-hydroxyacyl-CoA dehydrogenase activity; NAD binding; acetyl-CoA C-
 acetyltransferase activity; enoyl-CoA hydratase activity; fatty-acyl-CoA binding; long-
 chain-3-hydroxyacyl-CoA dehydrogenase activity; long-chain-enoil-CoA hydratase
 activity; lyase activity; oxidoreductase activity; protein binding;