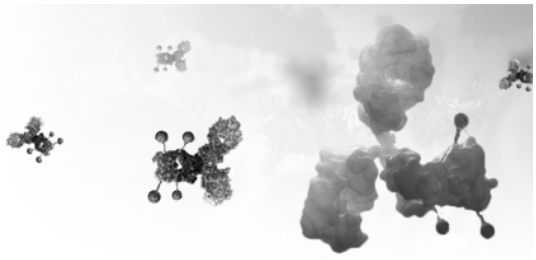

HADHA peptide (DAG-P0551)

This product is for research use only and is not intended for diagnostic use.

PRODUCT INFORMATION

Antigen 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. [provided by RefSeq, Jul 2008]
Purity	70 - 90% by HPLC.
Conjugate	Unconjugated
Sequence Similarities	In the N-terminal section; belongs to the enoyl-CoA hydratase/isomerase family.In the central section; belongs to the 3-hydroxyacyl-CoA dehydrogenase family.
Format	Liquid
Preservative	None
Storage	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles. Information available upon request.

GENE INFORMATION

Gene Name	HADHA hydroxyacyl-CoA dehydrogenase/3-ketoacyl-CoA thiolase/enoyl-CoA hydratase (trifunctional protein), alpha subunit [Homo sapiens (human)]
Official Symbol	HADHA
Synonyms	HADHA; hydroxyacyl-CoA dehydrogenase/3-ketoacyl-CoA thiolase/enoyl-CoA hydratase

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	(trifunctional protein), alpha subunit; GBP; ECHA; HADH; LCEH; MTPA; LCHAD; TP-ALPHA; trifunctional enzyme subunit alpha, mitochondrial; 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;
Entrez Gene ID	3030
mRNA Refseq	NM_000182.4
Protein Refseq	NP_000173.2
UniProt ID	E9KL44
Chromosome Location	2p23
Pathway	Acyl chain remodeling of CL, organism-specific biosystem; 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,
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-enoyl-CoA hydratase activity; protein bin

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