

Product datasheet for **TA318956**

HADHA Rabbit Polyclonal Antibody

Product data:

Product Type:	Primary Antibodies
Applications:	WB
Recommended Dilution:	WB: 0.5-4 ug/ml
Reactivity:	Human, Mouse, Rat
Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Immunogen:	Synthetic peptide surrounding amino acid 750 of human TFP1
Formulation:	100 µg (0.5 mg/ml) affinity purified rabbit polyclonal antibody in 1X phosphate-buffered saline (PBS) containing 30% glycerol, 0.5% BSA, and 0.01% thimerosal.
Concentration:	lot specific
Purification:	Affinity purified
Conjugation:	Unconjugated
Storage:	Store at -20°C as received.
Stability:	Stable for 12 months from date of receipt.
Gene Name:	hydroxyacyl-CoA dehydrogenase/3-ketoacyl-CoA thiolase/enoyl-CoA hydratase (trifunctional protein), alpha subunit
Database Link:	NP_000173 Entrez Gene 97212 Mouse Entrez Gene 170670 Rat Entrez Gene 3030 Human P40939
Background:	Mitochondrial Trifunctional Protein (TFP) is a multienzyme complex of the beta-oxidation cycle. TFP deficiency is a clinically heterogeneous disorder with phenotypes of different severity. The spectrum of diseases range from severe neonatal/infantile cardiomyopathy and early death to mild chronic progressive sensorimotor poly-neuropathy with episodic rhabdomyolysis. Human TFP is an octomer composed of four alpha-subunits and four beta-subunits. Mutations in either subunits may result in general TFP deficiency with reduced activity of all enzymes.
Synonyms:	ECHA; GBP; HADH; LCEH; LCHAD; MTPA; TP-ALPHA

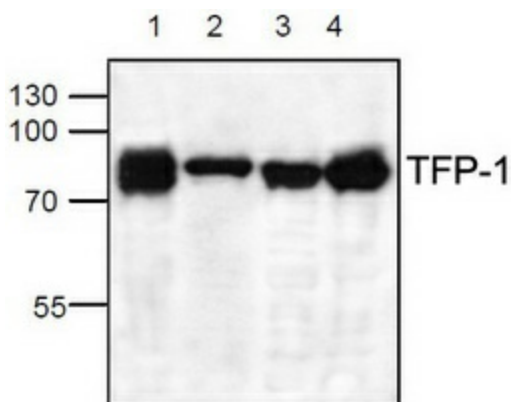


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Protein Families: Druggable Genome

Protein Pathways: beta-Alanine metabolism, Biosynthesis of unsaturated fatty acids, Butanoate metabolism, Fatty acid elongation in mitochondria, Fatty acid metabolism, Limonene and pinene degradation, Lysine degradation, Metabolic pathways, Propanoate metabolism, Tryptophan metabolism, Valine, leucine and isoleucine degradation

Product images:



Western blot analysis of TFP-1 expression with lysate from Jurkat cells (Lane 1, 2), 3T3 cells (Lane 3) and rat kidney (Lane 4).