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TFP1/HADHA Antibody
Rabbit Polyclonal Antibody
Catalog # ABV10552

Specification

TFP1/HADHA Antibody - Product Information

Application	**WB**
Primary Accession	P40939
Reactivity	**Human, Mouse, Rat**
Host	**Rabbit**
Clonality	**Polyclonal**
Isotype	**Rabbit IgG**
Calculated MW	**83000**

TFP1/HADHA Antibody - Additional Information

Gene ID 3030

Application & Usage	**Western blotting (0.5-4 µg/ml). However, the optimal concentrations should be determined individually. The antibody recognizes 84 kDa TFP1 from samples of human, mouse and rat origins. Reactivity to other species has not been tested.**

Other Names
Mitochondrial Trifunctional Protein

Target/Specificity
TFP1

Antibody Form
Liquid

Appearance
Colorless liquid

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.

Handling
The antibody solution should be gently mixed before use.

Reconstitution & Storage
-20 °C

Background Descriptions

Precautions

TFP1/HADHA Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

TFP1/HADHA Antibody - Protein Information

Name HADHA

Synonyms HADH

Function
Mitochondrial trifunctional enzyme catalyzes the last three of the four reactions of the mitochondrial beta-oxidation pathway (PubMed:8135828, PubMed:1550553, PubMed:29915090, PubMed:30850536). The mitochondrial beta-oxidation pathway is the major energy-producing process in tissues and is performed through four consecutive reactions breaking down fatty acids into acetyl-CoA (PubMed:29915090). Among the enzymes involved in this pathway, the trifunctional enzyme exhibits specificity for long-chain fatty acids (PubMed:30850536). Mitochondrial trifunctional enzyme is a heterotetrameric complex composed of two proteins, the trifunctional enzyme subunit alpha/HADHA described here carries the 2,3-enoyl-CoA hydratase and the 3-hydroxyacyl-CoA dehydrogenase activities while the trifunctional enzyme subunit beta/HADHB bears the 3-ketoacyl-CoA thiolase activity (PubMed:8135828, PubMed:29915090, PubMed:30850536). Independently of the subunit beta, the trifunctional enzyme subunit alpha/HADHA also has a monolysocardiolipin acyltransferase activity (PubMed:23152787). It acylates monolysocardiolipin into cardiolipin, a major mitochondrial membrane phospholipid which plays a key role in apoptosis and supports mitochondrial respiratory chain complexes in the generation of ATP (PubMed:23152787). Allows the acylation of monolysocardiolipin with different acyl-CoA substrates including oleoyl-CoA for which it displays the highest activity (PubMed:23152787).

Cellular Location
Mitochondrion. Mitochondrion inner membrane Note=Protein stability and association with mitochondrion inner membrane do not require HADHB.

TFP1/HADHA Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety

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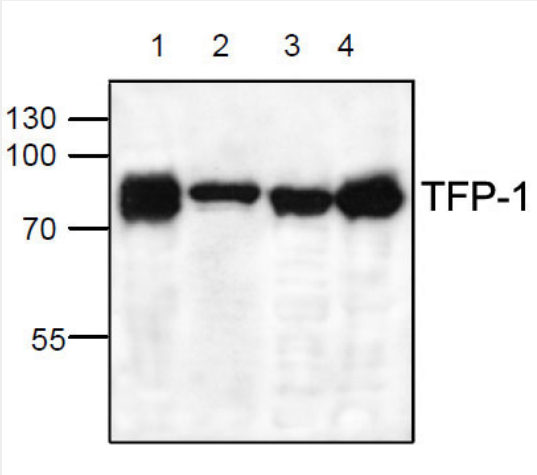
- Cell Culture

TFP1/HADHA Antibody - 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).

TFP1/HADHA Antibody - 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.