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Tafazzin Rabbit pAb

Catalog # AP99017

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	Q16635
Other Accession	Q16635
Predicted	Rat, Human, Mouse, Dog, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	30203
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Tafazzin
Epitope Specificity	201-292/292
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the taffazin family.
DISEASE	Barth syndrome (BTHS) [MIM:302060]: An X-linked disease characterized by dilated cardiomyopathy with endocardial fibroelastosis, a predominantly proximal skeletal myopathy, growth retardation, neutropenia, and organic aciduria, particularly excess of 3-methylglutaconic acid. Additional features include hypertrophic cardiomyopathy, isolated left ventricular non-compaction, ventricular arrhythmia, motor delay, poor appetite, fatigue and exercise intolerance, hypoglycemia, lactic acidosis, hyperammonemia, and dramatic late catch-up growth after growth delay throughout childhood. Note=The disease is caused by mutations affecting the gene represented in this entry.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes a protein that is expressed at high levels in cardiac and skeletal muscle. Mutations in this gene have been associated with a number of clinical disorders including Barth syndrome, dilated cardiomyopathy (DCM), hypertrophic DCM, endocardial fibroelastosis, and left ventricular noncompaction (LVNC). Multiple transcript variants encoding different isoforms have been described. A long form and a short form of each of these isoforms is produced; the short form lacks a hydrophobic leader sequence and may exist as a cytoplasmic protein rather than being membrane-bound. Other alternatively spliced transcripts have been described but the full-length nature of all these transcripts is not known. [provided by RefSeq, Jul 2008].

Additional Information

Gene ID	6901
Other Names	TAZ_HUMAN; TAFAZZIN; Taz; Protein G4.5; 2.3.1.-; EFE2; G4.5;

Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	TAFAZZIN (HGNC:11577)
Function	Acyltransferase required to remodel newly synthesized phospholipid cardiolipin (1',3'-bis-[1,2-diacyl-sn-glycero-3-phospho]- glycerol or CL), a key component of the mitochondrial inner membrane, with tissue specific acyl chains necessary for adequate mitochondrial function (PubMed:12930833, PubMed:19164547, PubMed:19700766, PubMed:26908608, PubMed:33096711). Its role in cellular physiology is to improve mitochondrial performance (PubMed:32234310). CL is critical for the coassembly of lipids and proteins in mitochondrial membranes, for instance, remodeling of the acyl groups of CL in the mitochondrial inner membrane affects the assembly and stability of respiratory chain complex IV and its supercomplex forms (By similarity). Catalyzes the transacylation between phospholipids and lysophospholipids, with the highest rate being between phosphatidylcholine (1,2-diacyl-sn-glycero- 3-phosphocholine or PC) and CL. Catalyzes both 1-acyl-sn-glycero-3- phosphocholine (lysophosphatidylcholine or LPC) reacylation and PC-CL transacylation, that means, it exchanges acyl groups between CL and PC by a combination of forward and reverse transacylations. Also catalyzes transacylations between other phospholipids such as phosphatidylethanolamine (1,2-diacyl-sn-glycero-3-phosphoethanolamine or PE) and CL, between PC and PE, and between PC and phosphatidate (1,2-diacyl-sn-glycero-3-phosphate or PA), although at lower rate. Not regiospecific, it transfers acyl groups into any of the sn-1 and sn-2 positions of the monolysocardiolipin (MLCL), which is an important prerequisite for uniformity and symmetry in CL acyl distribution. Cannot transacylate dilysocardiolipin (DLCL), thus, the role of MLCL is limited to that of an acyl acceptor. CoA-independent, it can reshuffle molecular species within a single phospholipid class. Redistributes fatty acids between MLCL, CL, and other lipids, which prolongs the half-life of CL. Its action is completely reversible, which allows for cyclic changes, such as fission and fusion or bending and flattening of the membrane. Hence, by contributing to the flexibility of the lipid composition, it plays an important role in the dynamics of mitochondria membranes. Essential for the final stage of spermatogenesis, spermatid individualization (By similarity). Required for the initiation of mitophagy (PubMed:33096711). Required to ensure progression of spermatocytes through meiosis (By similarity). Exon 7 of human tafazzin is essential for catalysis (PubMed:19700766).
Cellular Location	Mitochondrion outer membrane; Peripheral membrane protein; Intermembrane side. Mitochondrion inner membrane; Peripheral membrane protein; Intermembrane side [Isoform 2]: Cytoplasm. [Isoform 5]: Mitochondrion membrane [Isoform 7]: Mitochondrion membrane [Isoform 9]: Cytoplasm.
Tissue Location	High levels in cardiac and skeletal muscle. Up to 10 isoforms can be present in different amounts in different tissues Most isoforms are ubiquitous. Isoforms that lack the N-terminus are found in leukocytes and fibroblasts, but not in heart and skeletal muscle. Some forms appear restricted to cardiac and skeletal muscle or to leukocytes

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