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DNAJC19 Rabbit pAb

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Catalog # AP55552

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q96DA6
Reactivity	Mouse
Predicted	Human, Rat, Dog, Pig, Horse, Rabbit, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	12499
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human DNAJC19
Epitope Specificity	21-116/116
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Mitochondrion inner membrane.
SIMILARITY	Belongs to the TIM14 family. Contains 1 J domain.
DISEASE	Defects in DNAJC19 are the cause of 3-methylglutaconic aciduria type 5 (MGA5) [MIM:610198]; also known as dilated cardiomyopathy with ataxia (DCMA). MGA5 is an autosomal recessive disorder characterized by early-onset dilated cardiomyopathy, growth failure, cerebellar ataxia causing significant motor delays, testicular dysgenesis, growth failure, and significant increases in urine organic acids, particularly 3-methylglutaconic acid and 3-methylglutaric acid.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The protein encoded by this gene is thought to be part of a complex involved in the ATP-dependent transport of transit peptide-containing proteins from the inner cell membrane to the mitochondrial matrix. Defects in this gene are a cause of 3-methylglutaconic aciduria type 5 (MGA5), also known as dilated cardiomyopathy with ataxia (DCMA). Alternative splicing of this gene results in multiple transcript variants. Related pseudogenes have been identified on chromosomes 1, 2, 6, 10, 14 and 19. [provided by RefSeq, Jan 2012]

Additional Information

Gene ID	131118
Other Names	Mitochondrial import inner membrane translocase subunit TIM14, DnaJ homolog subfamily C member 19, DNAJC19, TIM14, TIMM14
Target/Specificity	Ubiquitously expressed.

Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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	DNAJC19
Synonyms	TIM14, TIMM14
Function	Mitochondrial co-chaperone which forms a complex with prohibitins to regulate cardiolipin remodeling (By similarity). May be a component of the PAM complex, a complex required for the translocation of transit peptide-containing proteins from the inner membrane into the mitochondrial matrix in an ATP-dependent manner. May act as a co-chaperone that stimulate the ATP-dependent activity (By similarity).
Cellular Location	Mitochondrion inner membrane; Single-pass membrane protein; Matrix side {ECO:0000250|UniProtKB:Q9CQV7}
Tissue Location	Ubiquitously expressed.

Background

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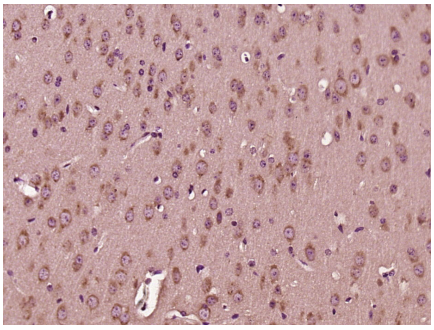
Images



Paraformaldehyde-fixed, paraffin embedded (Mouse brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (DNAJC19) Polyclonal Antibody, Unconjugated (AP55552) at 1:500 overnight at 4°C, followed by a conjugated secondary (sp-0023) for 20 minutes and DAB staining.