10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

TWNK Antibody (C-term)
Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP12799b

Specification

TWNK Antibody (C-term) - Product Information

Application	**WB, IHC-P,E**
Primary Accession	Q96RR1
Other Accession	NP_001157284.1, NP_068602.2
Reactivity	**Human**
Host	**Rabbit**
Clonality	**Polyclonal**
Isotype	**Rabbit IgG**
Antigen Region	**656-684**

TWNK Antibody (C-term) - Additional Information

Gene ID 56652

Other Names
Twinkle protein, mitochondrial, Progressive external ophthalmoplegia 1 protein, T7 gp4-like protein with intramitochondrial nucleoid localization, T7-like mitochondrial DNA helicase, PEO1, C10orf2

Target/Specificity
This C10orf2 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 656-684 amino acids from the C-terminal region of human C10orf2.

Dilution
WB~~1:1000
IHC-P~~1:10~50
E~~Use at an assay dependent concentration.

Format
Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.

Storage
Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions
TWNK Antibody (C-term) is for research use only and not for use in diagnostic or therapeutic procedures.

TWNK Antibody (C-term) - Protein Information

Name TWNK (HGNC:1160)

Synonyms C10orf2, PEO1

Function [Isoform 1]: Mitochondrial helicase involved in mtDNA replication and repair (PubMed:12975372, PubMed:15167897, PubMed:17324440, PubMed:18039713, PubMed:18971204, PubMed:25824949, PubMed:26887820, PubMed:27226550). Might have a role in mtDNA repair (PubMed:27226550). Has DNA strand separation activity needed to form a processive replication fork for leading strand synthesis which is catalyzed by the formation of a replisome complex with POLG and mtSDB (PubMed:12975372, PubMed:15167897, PubMed:18039713, PubMed:22383523, PubMed:26887820, PubMed:27226550). Preferentially unwinds DNA substrates with pre-existing 5'-and 3'- single-stranded tails but is also active on a 5'-flap substrate (PubMed:12975372, PubMed:15167897, PubMed:18039713, PubMed:22383523, PubMed:26887820, PubMed:27226550). Can dissociate the invading strand of immobile or mobile D-loop DNA structures irrespective of the single strand polarity of the third strand (PubMed:27226550). In addition to its DNA strand separation activity, also has DNA strand annealing, DNA strand-exchange and DNA branch migration activities (PubMed:22383523, PubMed:26887820, PubMed:27226550).

Cellular Location
Mitochondrion matrix, mitochondrion nucleoid Mitochondrion inner membrane; Peripheral membrane protein. Note=Colocalizes with mtDNA in mitochondrial nucleoids, a nucleoproteins complex consisting of a number of copies of proteins associated with mtDNA, probably involved in mtDNA maintenance and expression (PubMed:11431692) Associates with phospholipid membranes via electrostatic binding (By similarity). Preferentially associates with membranes enriched with cardiolipin, a lipid abundant in the mitochondrial inner membrane (PubMed:34950192). ATPase and helicase activity is enhanced by binding to lipid membranes (PubMed:34950192). {ECO:0000250|UniProtKB:Q9VL76, ECO:0000269|PubMed:11431692, ECO:0000269|PubMed:34950192}

Tissue Location
High relative levels in skeletal muscle, testis and pancreas. Lower levels of expression in the heart, brain, placenta, lung, liver, kidney, spleen, thymus, prostate, ovary, small intestine, colon and leukocytes. Expression is coregulated with MRPL43

TWNK Antibody (C-term) - 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
- Cell Culture

TWNK Antibody (C-term) - Images

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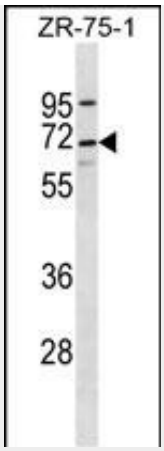
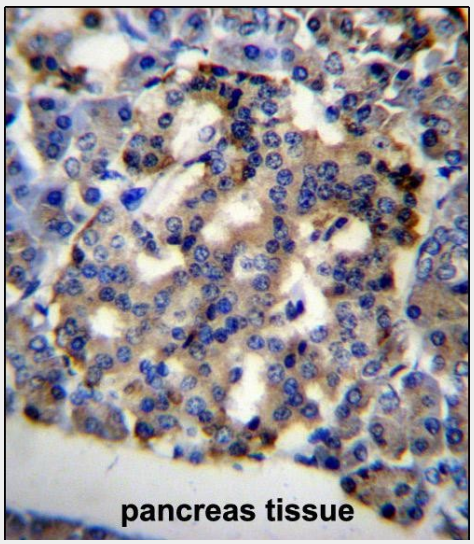


C10orf2 Antibody (C-term) (Cat. #AP12799b) western blot analysis in ZR-75-1 cell line lysates (35ug/lane).This demonstrates the C10orf2 antibody detected the C10orf2 protein (arrow).



C10orf2 Antibody (C-term) (Cat. #AP12799b)immunohistochemistry analysis in formalin fixed and paraffin embedded human pancreas tissue followed by peroxidase conjugation of the secondary antibody and DAB staining.This data demonstrates the use of C10orf2 Antibody (C-term) for immunohistochemistry. Clinical relevance has not been evaluated.

TWNK Antibody (C-term) - Background

This gene encodes a hexameric DNA helicase which unwinds short stretches of double-stranded DNA in the 5' to 3' direction and, along with mitochondrial single-stranded DNA binding protein and mtDNA polymerase gamma, is thought to play a key role in mtDNA replication. The protein localizes to the mitochondrial matrix and mitochondrial nucleoids. Mutations in this gene cause infantile onset spinocerebellar ataxia (IOSCA) and progressive external ophthalmoplegia (PEO) and are also associated with several mitochondrial depletion syndromes. Alternative splicing results in multiple transcript variants encoding distinct isoforms.

TWNK Antibody (C-term) - References

Longley, M.J., et al. J. Biol. Chem. 285(39):29690-29702(2010)
Wang, W., et al. Nucleic Acids Res. (2010) In press :

Fratter, C., et al. Neurology 74(20):1619-1626(2010)
Kruger, J., et al. Mol Neurodegener 5, 8 (2010) :
Bohlega, S., et al. Neuromuscul. Disord. 19(12):845-848(2009)