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C10orf2 Antibody (C-term) Blocking peptide
Synthetic peptide
Catalog # BP12799b

Specification

C10orf2 Antibody (C-term) Blocking peptide - Product Information

Primary Accession Q96RR1

C10orf2 Antibody (C-term) Blocking peptide - 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

Format
Peptides are lyophilized in a solid powder format. Peptides can be reconstituted in solution using the appropriate buffer as needed.

Storage
Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C.

Precautions
This product is for research use only. Not for use in diagnostic or therapeutic procedures.

C10orf2 Antibody (C-term) Blocking peptide - 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:<a href="http://www.uniprot.org/citations/18039713"

target="_blank">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

C10orf2 Antibody (C-term) Blocking peptide - Protocols

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

- Blocking Peptides

C10orf2 Antibody (C-term) Blocking peptide - Images

C10orf2 Antibody (C-term) Blocking peptide - Background

This gene encodes a hexameric DNA helicase which unwindsshort stretches of double-stranded DNA in the 5' to 3' directionand, along with mitochondrial single-stranded DNA binding proteinand mtDNA polymerase gamma, is thought to play a key role in mtDNAreplication. The protein localizes to the mitochondrial matrix andmitochondrial nucleoids. Mutations in this gene cause infantileonset spinocerebellar ataxia (IOSCA) and progressive externalophthalmoplegia (PEO) and are also associated with severalmitochondrial depletion syndromes. Alternative splicing results inmultiple transcript variants encoding distinct isoforms.

C10orf2 Antibody (C-term) Blocking peptide - 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)