

MIA40 Antibody (C-term) Blocking peptide
Synthetic peptide
Catalog # BP11528b**Specification**

MIA40 Antibody (C-term) Blocking peptide - Product InformationPrimary Accession [Q8N4Q1](#)**MIA40 Antibody (C-term) Blocking peptide - Additional Information****Gene ID** 131474**Other Names**Mitochondrial intermembrane space import and assembly protein 40,
Coiled-coil-helix-coiled-coil-helix domain-containing protein 4, CHCHD4, MIA40**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.

MIA40 Antibody (C-term) Blocking peptide - Protein Information**Name** CHCHD4**Synonyms** MIA40**Function**

Central component of a redox-sensitive mitochondrial intermembrane space import machinery which is required for the biogenesis of respiratory chain complexes (PubMed: 26004228). Functions as chaperone and catalyzes the formation of disulfide bonds in substrate proteins, such as COX17, COX19, MICU1 and COA7 (PubMed: 16185709, PubMed: 26387864, PubMed: 19182799, PubMed: 21059946, PubMed: 23186364, PubMed: 23676665, PubMed: 30885959). Required for the import and folding of small cysteine-containing proteins (small Tim) in the mitochondrial intermembrane space (IMS). Required for the import of COA7 in the IMS (PubMed: 30885959). Precursor proteins to be imported into the IMS are translocated

in their reduced form into the mitochondria. The oxidized form of CHCHD4/MIA40 forms a transient intermolecular disulfide bridge with the reduced precursor protein, resulting in oxidation of the precursor protein that now contains an intramolecular disulfide bond and is able to undergo folding in the IMS (PubMed:[16185709](http://www.uniprot.org/citations/16185709) PubMed:[19182799](http://www.uniprot.org/citations/19182799) PubMed:[21059946](http://www.uniprot.org/citations/21059946) PubMed:[23676665](http://www.uniprot.org/citations/23676665)). Reduced CHCHD4/MIA40 is then reoxidized by GFER/ERV1 via a disulfide relay system (PubMed:[23186364](http://www.uniprot.org/citations/23186364)). Mediates formation of disulfide bond in MICU1 in the IMS, promoting formation of the MICU1-MICU2 heterodimer that regulates mitochondrial calcium uptake (PubMed:[26387864](http://www.uniprot.org/citations/26387864)).

Cellular Location

Mitochondrion intermembrane space

Tissue Location

Expressed in all tissues tested, suggesting an ubiquitous expression.

MIA40 Antibody (C-term) Blocking peptide - Protocols

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

- [Blocking Peptides](#)

MIA40 Antibody (C-term) Blocking peptide - Images

MIA40 Antibody (C-term) Blocking peptide - Background

CHCHD4, a component of human mitochondria, belongs to a protein family whose members share 6 highly conserved cysteine residues constituting a -CXC-CX(9)C-CX(9)C- motif in the C terminus (Hofmann et al., 2005 [PubMed 16185709]).

MIA40 Antibody (C-term) Blocking peptide - References

Daithankar, V.N., et al. Biochemistry 48(22):4828-4837(2009) Chacinska, A., et al. J. Biol. Chem. 283(44):29723-29729(2008) Terziyska, N., et al. FEBS Lett. 581(6):1098-1102(2007) Hofmann, S., et al. J. Mol. Biol. 353(3):517-528(2005)