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MIA40 Antibody (C-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP11528b

Product Information

Application	WB, IHC-P, E
Primary Accession	Q8N4Q1
Other Accession	NP_001091972.1, NP_653237.1
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB29595
Calculated MW	15996
Antigen Region	101-129

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
Target/Specificity	This MIA40 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 101-129 amino acids from the C-terminal region of human MIA40.
Dilution	WB~~1:1000 IHC-P~~1:100~500 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	MIA40 Antibody (C-term) is for research use only and not for use in diagnostic or therapeutic procedures.

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 a chaperone and catalyzes the formation of disulfide bonds in substrate proteins, such as COA7, COX17, COX19, MICU1 and TIMCC/FAM136A (PubMed:16185709, PubMed:19182799, PubMed:21059946, PubMed:23186364, PubMed:23676665, PubMed:26387864, PubMed:30885959, PubMed:41109133). 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, PubMed:19182799, PubMed:21059946, PubMed:23676665). Reduced CHCHD4/MIA40 is then reoxidized by GFER/ERV1 via a disulfide relay system (PubMed: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).
Cellular Location	Mitochondrion intermembrane space
Tissue Location	Expressed in all tissues tested, suggesting an ubiquitous expression.

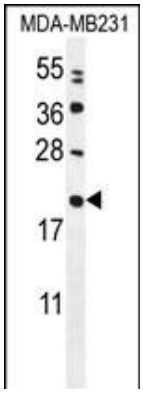
Background

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]).

References

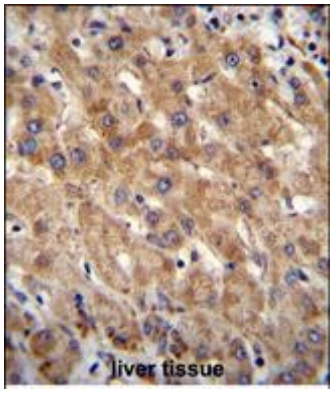
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Chacinska, A., et al. J. Biol. Chem. 283(44):29723-29729(2008)
Terziyska, N., et al. FEBS Lett. 581(6):1098-1102(2007)
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Images



MIA40 Antibody (C-term) (Cat. #AP11528b) western blot analysis in MDA-MB231 cell line lysates (35ug/lane).This demonstrates the MIA40 antibody detected the MIA40 protein (arrow).

MIA40 Antibody (C-term) (Cat. #AP11528b)immunohistochemistry analysis in formalin fixed and paraffin embedded human liver tissue followed



by peroxidase conjugation of the secondary antibody and DAB staining.This data demonstrates the use of MIA40 Antibody (C-term) for immunohistochemistry. Clinical relevance has not been evaluated.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.