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MOCS1 Rabbit pAb

Catalog # AP99480

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	Q9NZB8
Other Accession	Q9NZB8
Predicted	Rat, Pig, Human, Mouse, Rabbit, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	70105
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human MOCS1
Epitope Specificity	21-120/636
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	In the C-terminal section; belongs to the moaC family. In the N-terminal section; belongs to the moaA/nifB/pqqE family.
Post-translational modifications	Isoform MOCS1A, isoform 2 and isoform 3 are probably thiocarboxylated at their C-terminus. Thiocarboxylation probably plays a central role in molybdenum cofactor biosynthesis, since mutagenesis of the last 2 Gly residues of isoform MOCS1A abolishes the catalytic activity of the enzyme. Thiocarboxylation is absent in isoform MOCS1B, which lacks the C-terminal Gly residue.
DISEASE	Defects in MOCS1 are the cause of molybdenum cofactor deficiency type A (MOCOD type A) [MIM:252150]; an autosomal recessive disease which leads to the pleiotropic loss of all molybdoenzyme activities and is characterized by severe neurological damage, neonatal seizures and early childhood death.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Molybdenum cofactor biosynthesis is a conserved pathway leading to the biological activation of molybdenum. The protein encoded by this gene is involved in this pathway. This gene was originally thought to produce a bicistronic mRNA with the potential to produce two proteins (MOCS1A and MOCS1B) from adjacent open reading frames. However, only the first open reading frame (MOCS1A) has been found to encode a protein from the putative bicistronic mRNA, whereas additional splice variants, whose full-length natures have yet to be determined, are likely to produce a fusion between the two open reading frames. This gene is defective in patients with molybdenum cofactor deficiency, type A. A related pseudogene has been identified on chromosome 16. [provided by RefSeq, Jan 2010]

Additional Information

Gene ID	4337

Other Names	MIG11; MOCOD; MOCS1A; MOCS1B; 3110045D15Rik; MOCS1_BOVIN; MOCS1; MOCS1_HUMAN; Cell migration-inducing gene 11 protein; Molybdenum cofactor synthesis-step 1 protein A-B; MOCS1_MOUSE; molybdenum cofactor synthesis 1
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
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	MOCS1
Function	Isoform MOCS1A and isoform MOCS1B probably form a complex that catalyzes the conversion of 5'-GTP to cyclic pyranopterin monophosphate (cPMP) (PubMed:11891227, PubMed:23627491, PubMed:29368224, PubMed:31996372). MOCS1A catalyzes the cyclization of GTP to (8S)-3',8-cyclo-7,8-dihydroguanosine 5'-triphosphate and MOCS1B catalyzes the subsequent conversion of (8S)-3',8-cyclo-7,8- dihydroguanosine 5'-triphosphate to cPMP (PubMed:11891227, PubMed:23627491, PubMed:29368224, PubMed:31996372).
Cellular Location	[Isoform MOCS1A]: Mitochondrion matrix [Isoform 9]: Mitochondrion matrix [Isoform 3]: Cytoplasm. Note=Found in aggregates not co- localized with mitochondria.
Tissue Location	Isoform MOCS1A and isoform 2 are widely expressed.

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