

钼辅因子合成蛋白1抗体

产品货号：mlR17700

英文名称：MOCS1

中文名称：钼辅因子合成蛋白1抗体

别 名：Cell migration-inducing gene 11 protein; MIG11; MOCOD; Mocs1; MOCS1_HUMAN; Molybdenum cofactor biosynthesis protein 1; Molybdenum cofactor biosynthesis protein A; Molybdenum cofactor biosynthesis protein C; Molybdenum cofactor synthesis 1; Molybdenum cofactor synthesis-step 1 protein A-B.

研究领域：细胞生物 神经生物学 信号转导 新陈代谢

抗体来源：Rabbit

克隆类型：Polyclonal

交叉反应：Human, Mouse, Rat, Pig, Horse, Rabbit,

产品应用：ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分子量：70kDa

细胞定位：细胞核 细胞浆

性 状：Lyophilized or Liquid

浓 度：1mg/ml

免疫原：KLH conjugated synthetic peptide derived from human MOCS1:21-120/636

亚 型：IgG

纯化方法：affinity purified by Protein A

储存液 : 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件 : Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.

PubMed : PubMed

产品介绍 : 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]

Function:

Isoform MOCS1A and isoform MOCS1B probably form a complex that catalyzes the conversion of a guanosine derivative to precursor Z during molybdenum cofactor biosynthesis.

Tissue Specificity:

Isoform MOCS1A and isoform 2 are widely expressed.

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.

Similarity:

In the C-terminal section; belongs to the moaC family.

In the N-terminal section; belongs to the moaA/nifB/pqqE family.

SWISS:

Q9NZB8



Gene ID:

4337

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.