

MOCS2 (G-9): sc-377343

BACKGROUND

MOCS2 (molybdopterin synthase) is a heterotetrameric synthase composed of two small (MOCS2A) and two large (MOCS2B) subunits. The small and large subunits are both encoded by a single bicistronic mRNA, with the open reading frames overlapping by 77 nucleotides. MOCS2 functions in the second step of the synthesis of molybdenum cofactor or molybdopterin (MPT). It catalyzes the formation of MPT from precursor Z by incorporating a dithiolene functional group. The C-terminus of the small subunit of MOCS2 acts as the sulfur donor for the synthesis of this functional group. MPT is inserted into molybdoenzymes and is required for the proper function of aldehyde oxidase, xanthine dehydrogenase and sulphite oxidase enzymes. Mutations in the gene encoding MOCS2 can lead to molybdenum cofactor deficiency and can result in early childhood death.

REFERENCES

1. Reiss, J., et al. 1999. Human molybdopterin synthase gene: genomic structure and mutations in molybdenum cofactor deficiency type B. *Am. J. Hum. Genet.* 64: 706-711.
2. Rudolph, M.J., et al. 2003. Structural studies of molybdopterin synthase provide insights into its catalytic mechanism. *J. Biol. Chem.* 278: 14514-14522.
3. Leimkuhler, S., et al. 2003. Mechanistic studies of human molybdopterin synthase reaction and characterization of mutants identified in group B patients of molybdenum cofactor deficiency. *J. Biol. Chem.* 278: 26127-26134.
4. Leimkuhler, S., et al. 2005. Ten novel mutations in the molybdenum cofactor genes MOCS1 and MOCS2 and *in vitro* characterization of a MOCS2 mutation that abolishes the binding ability of molybdopterin synthase. *Hum. Genet.* 117: 565-570.
5. Singh, S., et al. 2005. Three-dimensional structure of the AAH26994.1 protein from *Mus musculus*, a putative eukaryotic Urm1. *Protein Sci.* 14: 2095-2102.
6. Turpaev, K., et al. 2005. Analysis of differentially expressed genes in nitric oxide-exposed human monocytic cells. *Free Radic. Biol. Med.* 38: 1392-1400.
7. Suzuki, M., et al. 2006. The maize viviparous15 locus encodes the molybdopterin synthase small subunit. *Plant J.* 45: 264-274.
8. Hahnwald, R., et al. 2006. A novel MOCS2 mutation reveals coordinated expression of the small and large subunit of molybdopterinsynthase. *Mol. Genet. Metab.* 89: 210-213.
9. Per, H., et al. 2007. Molybdenum cofactor deficiency: clinical features in a Turkish patient. *Brain Dev.* 29: 365-368.

CHROMOSOMAL LOCATION

Genetic locus: MOCS2 (human) mapping to 5q11.2.

SOURCE

MOCS2 (G-9) is a mouse monoclonal antibody raised against amino acids 1-188 representing full length MOCS2 of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

MOCS2 (G-9) is recommended for detection of MOCS2 of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for MOCS2 siRNA (h): sc-72268, MOCS2 shRNA Plasmid (h): sc-72268-SH and MOCS2 shRNA (h) Lentiviral Particles: sc-72268-V.

Molecular Weight of MOCS2 large subunit: 21 kDa.

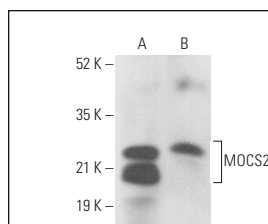
Molecular Weight of MOCS2 small subunit: 10 kDa.

Positive Controls: Jurkat whole cell lysate: sc-2204, Hep G2 cell lysate: sc-2227 or MOCS2 (h): 293T Lysate: sc-116247.

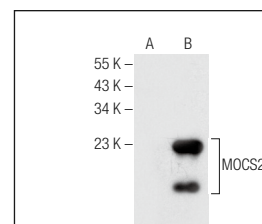
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



MOCS2 (G-9): sc-377343. Western blot analysis of MOCS2 expression in Jurkat (A) and Hep G2 (B) whole cell lysates.



MOCS2 (G-9): sc-377343. Western blot analysis of MOCS2 expression in non-transfected: sc-117752 (A) and human MOCS2 transfected: sc-116247 (B) 293T whole cell lysates.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.