SANTA CRUZ BIOTECHNOLOGY, INC.

MOCS2 (h): 293T Lysate: sc-116247

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. Leimkühler, 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. Hahnewald, 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.

PRODUCT

MOCS2 (h): 293T Lysate represents a lysate of human MOCS2 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

MOCS2 (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive MOCS2 antibodies. Recommended use: 10-20 µl per lane.

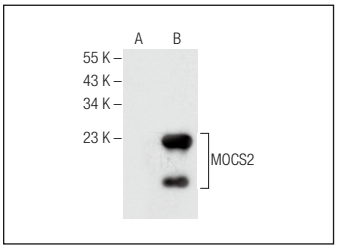
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

MOCS2 (G-9): sc-377343 is recommended as a positive control antibody for Western Blot analysis of enhanced human MOCS2 expression in MOCS2 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

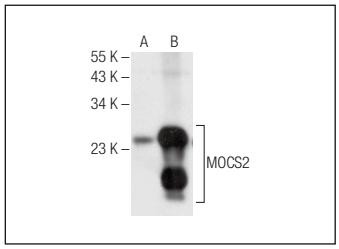
RECOMMENDED SUPPORT REAGENTS

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.

DATA



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.



MOCS2 (F-9): sc-377169. 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 -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.