

MAAI (H-4): sc-374404

BACKGROUND

In humans, maleylacetoacetate isomerase (MAAI, also known as GSTZ1-1) catalyzes the conversion of maleylacetoacetate to fumarylacetoacetate, the fifth step in the phenylalanine/phenylacetate degradation pathway. Deficiencies in other steps of this pathway cause metabolic diseases, including type I tyrosinemia and phenylketonuria. The glutathione S-transferases (GSTs) are found in all aerobic organisms and catalyze the conjugation of glutathione to a wide variety of electrophilic substrates. By sequence alignment and phylogenetic analysis, a new subgroup of GST-like proteins from human, *C. elegans*, and carnation were identified. Human MAAI is 38% and 49% identical to the carnation and *C. elegans* proteins, respectively. Recombinant human MAAI is a dimer. The enzyme exhibits limited activity with known GST substrates. Western blot analysis indicates that MAAI is most abundant in liver, with lower levels detected in skeletal muscle and brain. The gene which encodes MAAI maps to human chromosome 14q24.3.

REFERENCES

- Berger, R., et al. 1988. Tyrosinemia type Ib caused by maleylacetoacetate isomerase deficiency: a new enzyme defect. *Pediat. Res.* 23: 328A.
- Board, P.G., et al. 1997. ζ , a novel class of glutathione transferases in a range of species from plants to humans. *Biochem. J.* 328: 929-935.
- Blackburn, A.C., et al. 1998. Characterization and chromosome location of the gene GSTZ1 encoding the human ζ class glutathione transferase and maleylacetoacetate isomerase. *Cytogenet. Cell Genet.* 83: 109-114.
- Fernandez-Canon, J.M., et al. 1998. Characterization of a fungal maleylacetoacetate isomerase gene and identification of its human homologue. *J. Biol. Chem.* 273: 329-337.
- Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 603758. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>

CHROMOSOMAL LOCATION

Genetic locus: GSTZ1 (human) mapping to 14q24.3.

SOURCE

MAAI (H-4) is a mouse monoclonal antibody raised against amino acids 1-216 representing full length MAAI of human origin.

PRODUCT

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

STORAGE

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

PROTOCOLS

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

APPLICATIONS

MAAI (H-4) is recommended for detection of MAAI of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (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 MAAI siRNA (h): sc-40729, MAAI shRNA Plasmid (h): sc-40729-SH and MAAI shRNA (h) Lentiviral Particles: sc-40729-V.

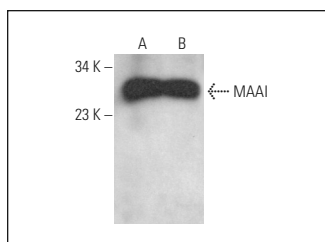
Molecular Weight of MAAI: 24 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227 or MCF7 whole cell lysate: sc-2206.

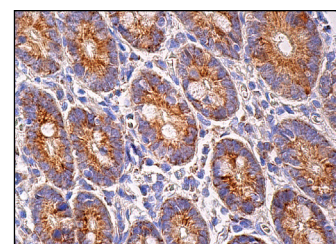
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ 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-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgG κ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



MAAI (H-4): sc-374404. Western blot analysis of MAAI expression in Hep G2 (A) and MCF7 (B) whole cell lysates.



MAAI (H-4): sc-374404. Immunoperoxidase staining of formalin fixed, paraffin-embedded human duodenum tissue showing cytoplasmic staining of glandular cells.

RESEARCH USE

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