



MAAI (m): 293T Lysate: sc-121475

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 composed of 24.2-kD subunits. 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

1. Berger, R., Michals, K., Galbraeth, J. and Matalon, R. 1988. Tyrosinemia type Ib caused by maleylacetoacetate isomerase deficiency: a new enzyme defect. *Pediatr. Res.* 23: 328A.
2. Board, P.G., Baker, R.T., Chelvanayagam, G. and Jermini, L.S. 1997. ζ , a novel class of glutathione transferases in a range of species from plants to humans. *Biochem. J.* 328: 929-935.
3. Blackburn, A.C., Woollatt, E., Sutherland, G.R. and Board, P.G. 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.
4. Fernandez-Canon, J.M. and Penalva, M.A. 1998. Characterization of a fungal maleylacetoacetate isomerase gene and identification of its human homologue. *J. Biol. Chem.* 273: 329-337.
5. 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 (mouse) mapping to 12 D2.

PRODUCT

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

APPLICATIONS

MAAI (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive MAAI 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.

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.