## 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 $24.2-\mathrm{kD}$ 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., et al. 1988. Tyrosinemia type lb caused by maleylacetoacetate isomerase deficiency: a new enzyme defect. Pediat. Res. 23 Suppl.: 328A.
2. Board, P.G., et al. 1997. $\zeta$, a novel class of glutathione transferases in a range of species from plants to humans. Biochem. J. 328: 929-935.
3. Blackburn, A.C., et al. 1998. Characterization and chromosome location of the gene GSTZ1 encoding the human $\zeta$ class glutathione transferase and maleylacetoacetate isomerase. Cytogenet. Cell Genet. 83: 109-114.
4. 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.
5. Online Mendelian Inheritance in Man, OMIM ${ }^{\top M}$. 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; Gstz1 (mouse) mapping to 12 D 2 .

## SOURCE

MAAI (FL-216) is a rabbit polyclonal antibody raised against amino acids $1-216$ representing full length MAAl of human origin.

## PRODUCT

Each vial contains $200 \mu \mathrm{ggG}$ in 1.0 ml of PBS with $<0.1 \%$ sodium azide and $0.1 \%$ gelatin.

## STORAGE

Store at $4^{\circ} \mathrm{C}$, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

## APPLICATIONS

MAAI (FL-216) is recommended for detection of MAAI of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [ $1-2 \mu \mathrm{~g}$ per 100-500 $\mu \mathrm{g}$ 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).

MAAI (FL-216) is also recommended for detection of MAAI in additional species, including equine, canine, bovine and porcine.
Suitable for use as control antibody for MAAI siRNA (h): sc-40729, MAAI siRNA (m): sc-40730, MAAI shRNA Plasmid (h): sc-40729-SH, MAAI shRNA Plasmid (m): sc-40730-SH, MAAI shRNA (h) Lentiviral Particles: sc-40729-V and MAAI shRNA (m) Lentiviral Particles: sc-40730-V.

Molecular Weight of MAAI: 24 kDa .
Positive Controls: human liver extract: sc-363766, mouse liver extract: sc-2256 or Hep G2 cell lysate: sc-2227.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat antirabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz ${ }^{\text {TM }}$ Mounting Medium: sc-24941.

## DATA



MAAI (FL-216): sc-32934. Western blot analysis of MAAI expression in human liver tissue extract.

## RESEARCH USE

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


