

MMAB (H-217): sc-98743

BACKGROUND

MMAB (methylmalonic aciduria (cobalamin deficiency) type B protein), also known as ATR or Cob(I)alamin adenosyltransferase, is a mitochondrial protein expressed in skeletal muscle and liver. MMAB belongs to the Cob(I)alamin adenosyltransferase family and plays an important role in adenosylcobalamin (AdoCbl) biosynthesis. More specifically, MMAB catalyzes the final step in the biosynthesis pathway: the conversion of vitamin B12 (also known as cobalamin) to AdoCbl. AdoCbl is an essential cofactor utilized by MUT, the mitochondrial methylmalonyl-CoA mutase that plays an important role in the catabolism of cholesterol, branched chain amino acids, odd-numbered fatty acids and other metabolites. Mutations in the gene encoding MMAB can result in methylmalonic aciduria type B (MMAB), also known as vitamin B12-responsive methylmalonicaciduria of cblB complementation type. The autosomal recessive MMAB disease is characterized by defective synthesis of AdoCbl.

REFERENCES

1. Johnson, C.L., et al. 2001. Functional genomic, biochemical and genetic characterization of the *Salmonella pduO* gene, an ATP: Cob(I)alamin adenosyltransferase gene. *J. Bacteriol.* 183: 1577-1584.
2. Dobson, C.M., et al. 2002. Identification of the gene responsible for the CblB complementation group of vitamin B12-dependent methylmalonic aciduria. *Hum. Mol. Genet.* 11: 3361-3369.
3. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 607568. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
4. Saridakis, V., et al. 2004. The structural basis for methylmalonic aciduria. The crystal structure of archaeal ATP: cobalamin adenosyltransferase. *J. Biol. Chem.* 279: 23646-23653.
5. Lerner-Ellis, J.P., et al. 2006. Mutation and biochemical analysis of patients belonging to the CblB complementation class of vitamin B12-dependent methylmalonic aciduria. *Mol. Genet. Metab.* 87: 219-225.

CHROMOSOMAL LOCATION

Genetic locus: MMAB (human) mapping to 12q24.11; Mmab (mouse) mapping to 5 F.

SOURCE

MMAB (H-217) is a rabbit polyclonal antibody raised against amino acids 34-250 mapping at the C-terminus of MMAB of human origin.

PRODUCT

Each vial contains 200 µg IgG 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.

APPLICATIONS

MMAB (H-217) is recommended for detection of MMAB of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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 MMAB siRNA (h): sc-75802, MMAB siRNA (m): sc-75803, MMAB shRNA Plasmid (h): sc-75802-SH, MMAB shRNA Plasmid (m): sc-75803-SH, MMAB shRNA (h) Lentiviral Particles: sc-75802-V and MMAB shRNA (m) Lentiviral Particles: sc-75803-V.

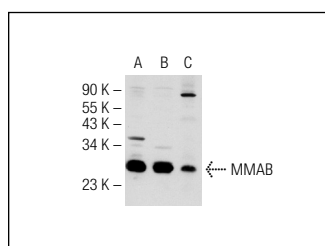
Molecular Weight of MMAB: 27 kDa.

Positive Controls: DU 145 cell lysate: sc-2268, Hep G2 cell lysate: sc-2227 or mouse skeletal muscle tissue.

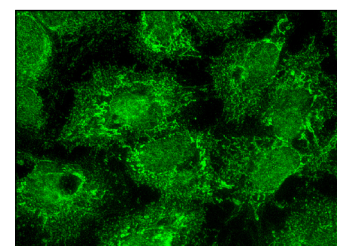
RECOMMENDED SECONDARY REAGENTS

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 anti-rabbit 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™ Mounting Medium: sc-24941.

DATA



MMAB (H-217): sc-98743. Western blot analysis of MMAB expression in DU 145 (A) and Hep G2 (B) whole cell lysates and mouse skeletal muscle tissue extract (C).



MMAB (H-217): sc-98743. Immunofluorescence staining of formalin-fixed Hep G2 cells showing mitochondria localization.

RESEARCH USE

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

MONOS
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Try **MMAB (G-3): sc-271424**, our highly recommended monoclonal alternative to MMAB (H-217).