SANTA CRUZ BIOTECHNOLOGY, INC.

MUT (D-1): sc-390978

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

MUT (methylmalonyl coenzyme A mutase), also known as MCM, is a 750 amino acid mitochondrial matrix protein that exists as a homodimer and belongs to the methylmalonyl-CoA mutase family. Induced by adenosylcobalamin (also known as coenzyme B12 or vitamin B12), MUT participates in the degradation of various amino acids, odd-chain fatty acids and cholesterol via propionyl-CoA (PCC) during the tricarboxylic acid cycle. Mutations in the gene encoding MUT, which is located on human chromsome 6, are the cause of methylmalonic aciduria type mut (MMAM), an often fatal disorder of organic acid metabolism that is characterized by lethargy, vomiting, failure to thrive, hypotonia, neurological deficit and early death. Two forms of MMAM exists: mut(o), which there is no detectable enzymatic activity and mut$^-$, which there is residual cobalamin-dependent activity.

REFERENCES

1. Wilkemeyer, M.F., et al. 1991. Differential diagnosis of mut and cbl methylmalonic aciduria by DNA-mediated gene transfer in primary fibroblasts. J. Clin. Invest. 87: 915-918.
2. Crane, A.M., et al. 1992. Cloning and expression of a mutant methylmalonyl coenzyme A mutase with altered cobalamin affinity that causes mut-methylmalonic aciduria. J. Clin. Invest. 89: 385-391.
3. Crane, A.M., et al. 1994. Clustering of mutations in methylmalonyl CoA mutase associated with mut-methylmalonic acidemia. Am. J. Hum. Genet. 55: 42-50.
4. Treacy, E., et al. 1996. Glutathione deficiency as a complication of methylmalonic acidemia: response to high doses of ascorbate. J. Pediatr. 129: 445-448.
5. Janata, J., et al. 1997. Expression and kinetic characterization of methylmalonyl-CoA mutase from patients with the mut- phenotype: evidence for naturally occurring interallelic complementation. Hum. Mol. Genet. 6: 1457-1464.
6. Ledley, F.D., et al. 1997. Mutations in mut methylmalonic acidemia: clinical and enzymatic correlations. Hum. Mutat. 9: 1-6.
7. Mücher, G., et al. 1998. Fine mapping of the autosomal recessive polycystic kidney disease locus (PKHD1) and the genes MUT, RDS, CSNK2 β, and GSTA1 at 6p21.1-p12. Genomics 48: 40-45.

CHROMOSOMAL LOCATION

Genetic locus: MUT (human) mapping to 6p12.3; Mut (mouse) mapping to 17 B2.

SOURCE

MUT (D-1) is a mouse monoclonal antibody raised against amino acids 451-750 mapping at the C-terminus of MUT of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

MUT (D-1) is recommended for detection of MUT of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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), 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 MUT siRNA (h): sc-95089, MUT siRNA (m): sc-149723, MUT shRNA Plasmid (h): sc-95089-SH, MUT shRNA Plasmid (m): sc-149723-SH, MUT shRNA (h) Lentiviral Particles: sc-95089-V and MUT shRNA (m) Lentiviral Particles: sc-149723-V.

Molecular Weight of mature MUT: 78 kDa.

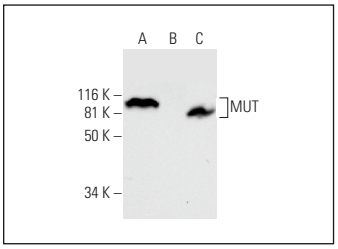
Molecular Weight of MUT precursor: 82 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227 or K-562 whole cell lysate: sc-2203.

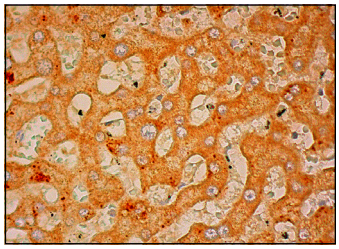
RECOMMENDED SUPPORT REAGENTS

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. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ 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-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



MUT (D-1): sc-390978. Western blot analysis of MUT expression in Hep G2 (**A**), NIH/3T3 (**B**) and K-562 (**C**) whole cell lysates. Note lack of reactivity with mouse MUT in lane **B**.



MUT (D-1): sc-390978. Immunoperoxidase staining of formalin fixed, paraffin-embedded human liver tissue showing cytoplasmic staining of hepatocytes.

STORAGE

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

RESEARCH USE

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