SANTA CRUZ BIOTECHNOLOGY, INC.

MUT (24): sc-136541

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 6p12.3, 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. and Rosenblatt, D.S. 1997. Mutations in MUT methylmalonic acidemia: clinical and enzymatic correlations. Hum. Mutat. 9: 1-6.

CHROMOSOMAL LOCATION

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

SOURCE

MUT (24) is a mouse monoclonal antibody raised against amino acids 1-102 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.

MUT (24) is available conjugated to agarose (sc-136541 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; and to HRP (sc-136541 HRP), 200 µg/ml, for WB, IHC(P) and ELISA.

APPLICATIONS

MUT (24) is recommended for detection of MUT of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) and immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)].

MUT (24) is also recommended for detection of MUT in additional species, including canine.

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.

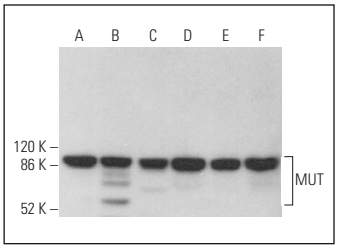
Molecular Weight of MUT precursor: 82 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, HEK293 whole cell lysate: sc-45136 or K-562 whole cell lysate: sc-2203.

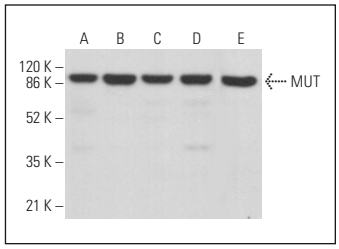
RECOMMENDED SUPPORT REAGENTS

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).

DATA



MUT (24): sc-136541. Western blot analysis of MUT expression in Hep G2 (**A**), Caki-1 (**B**), HL-60 (**C**), RAW 264.7 (**D**), 3T3-L1 (**E**) and C3H/10T1/2 (**F**) whole cell lysates.



MUT (24): sc-136541. Western blot analysis of MUT expression in HeLa (**A**), NIH/3T3 (**B**), K-562 (**C**), HEK293 (**D**) and A-431 (**E**) whole cell lysates. Detection reagent used: m-IgGκ BP-HRP: sc-516102.

SELECT PRODUCT CITATIONS

1. Costanzo, M., et al. 2018. Label-free quantitative proteomics in a methylmalonyl-CoA mutase-silenced neuroblastoma cell line. Int. J. Mol. Sci. 19 pii: E3580.

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.