SANTA CRUZ BIOTECHNOLOGY, INC.

MUT (D-13): sc-161117

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. and Ledley, F.D. 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.
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.
8. Kobayashi, A., et al. 2006. Three novel and six common mutations in 11 patients with methylmalonic acidemia. Pediatr. Int. 48: 1-4.
9. Hu, R., et al. 2009. Gene induction for the treatment of methylmalonic aciduria. J. Gene Med. 11: 361-369.

CHROMOSOMAL LOCATION

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

SOURCE

MUT (D-13) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of MUT 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.

Blocking peptide available for competition studies, sc-161117 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

MUT (D-13) 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), 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).

MUT (D-13) is also recommended for detection of MUT in additional species, including equine, canine, porcine and avian.

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 MUT precursor: 82 kDa.

Molecular Weight of mature MUT: 78 kDa.

Positive Controls: rat liver extract: sc-2395.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

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.