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MMACHC (B-5): sc-518236

BACKGROUND

MMACHC (methylmalonic aciduria and homocystinuria type C protein), also known as cblC, is a 282 amino acid widely expressed protein that may be involved in the binding and intracellular trafficking of cobalamin (vitamin B12). Defects in the gene encoding MMACHC are the cause of methylmalonic aciduria and homocystinuria type cblC, a disorder of cobalamin metabolism characterized by decreased levels of the coenzymes adenosylcobalamin (AdoCbl) and methylcobalamin (MeCbl). 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. MeCbl is an active coenzyme form of vitamin B-12 and is essential for cell growth and replication. Individuals affected by methylmalonic aciduria and homocystinuria type cblC experience negative developmental, hematologic, neurologic, metabolic, ophthalmologic, and dermatologic manifestations.

REFERENCES

1. Morel, C.F., et al. 2006. Combined methylmalonic aciduria and homocystinuria (cblC): phenotype-genotype correlations and ethnic-specific observations. *Mol. Genet. Metab.* 88: 315-321.
2. Tsai, A.C., et al. 2007. Late-onset combined homocystinuria and methylmalonic aciduria (cblC) and neuropsychiatric disturbance. *Am. J. Med. Genet. A* 143A: 2430-2434.
3. Nogueira, C., et al. 2008. Spectrum of MMACHC mutations in Italian and Portuguese patients with combined methylmalonic aciduria and homocystinuria, cblC type. *Mol. Genet. Metab.* 93: 475-480.
4. Kim, J., et al. 2008. Decyanation of vitamin B12 by a trafficking chaperone. *Proc. Natl. Acad. Sci. USA* 105: 14551-14554.
5. Richard, E., et al. 2009. Genetic and cellular studies of oxidative stress in methylmalonic aciduria (MMA) cobalamin deficiency type C (cblC) with homocystinuria (MMACHC). *Hum. Mutat.* 30: 1558-1566.

CHROMOSOMAL LOCATION

Genetic locus: MMACHC (human) mapping to 1p34.1.

SOURCE

MMACHC (B-5) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 100-119 of MMACHC of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

MMACHC (B-5) is available conjugated to agarose (sc-518236 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-518236 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-518236 PE), fluorescein (sc-518236 FITC), Alexa Fluor® 488 (sc-518236 AF488), Alexa Fluor® 546 (sc-518236 AF546), Alexa Fluor® 594 (sc-518236 AF594) or Alexa Fluor® 647 (sc-518236 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-518236 AF680) or Alexa Fluor® 790 (sc-518236 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

MMACHC (B-5) is recommended for detection of MMACHC of 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

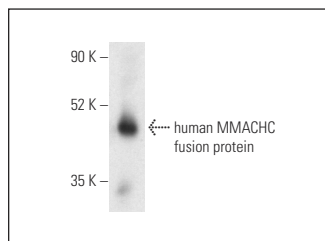
Suitable for use as control antibody for MMACHC siRNA (h): sc-88005, MMACHC shRNA Plasmid (h): sc-88005-SH and MMACHC shRNA (h) Lentiviral Particles: sc-88005-V.

Molecular Weight of MMACHC: 32 kDa.

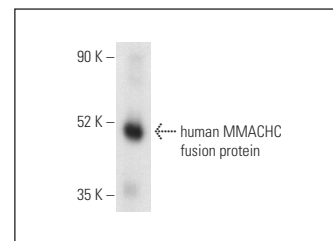
RECOMMENDED SUPPORT REAGENTS

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.

DATA



MMACHC (B-5): sc-518236. Western blot analysis of human recombinant MMACHC fusion protein. Detection reagent used: m-IgG₁ BP-HRP: sc-525408.



MMACHC (B-5): sc-518236. Western blot analysis of human recombinant MMACHC fusion protein. Detection reagent used: m-IgG Fc BP-HRP: sc-525409.

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.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.

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