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MMAA Antibody (N-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP9795a

Product Information

Application	WB, IHC-P, E
Primary Accession	Q8IVH4
Other Accession	Q8C7H1
Reactivity	Human, Mouse
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB24782
Calculated MW	46538
Antigen Region	56-84

Additional Information

Gene ID	166785
Other Names	Methylmalonic aciduria type A protein, mitochondrial, 36--, MMAA
Target/Specificity	This MMAA antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 56-84 amino acids from the N-terminal region of human MMAA.
Dilution	WB~~1:1000 IHC-P~~1:100~500 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	MMAA Antibody (N-term) is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	MMAA (HGNC:18871)
Function	GTPase, binds and hydrolyzes GTP (PubMed: 20876572, PubMed:21138732, PubMed:28497574, PubMed:28943303). Involved in intracellular vitamin B12 metabolism, mediates the transport of cobalamin (Cbl) into mitochondria for the final steps of adenosylcobalamin (AdoCbl) synthesis (PubMed:20876572,

	PubMed:28497574). Functions as a G-protein chaperone that assists AdoCbl cofactor delivery from MMAB to the methylmalonyl-CoA mutase (MMUT) (PubMed:20876572, PubMed:28497574). Plays a dual role as both a protectase and a reactivase for MMUT (PubMed:21138732, PubMed:28943303). Protects MMUT from progressive inactivation by oxidation by decreasing the rate of the formation of the oxidized inactive cofactor hydroxocobalamin (OH2Cbl) (PubMed:21138732, PubMed:28943303). Additionally acts a reactivase by promoting the replacement of OH2Cbl by the active cofactor AdoCbl, restoring the activity of MMUT in the presence and hydrolysis of GTP (PubMed:21138732, PubMed:28943303).
Cellular Location	Mitochondrion {ECO:0000269|PubMed:28943303, ECO:0000305}. Cytoplasm
Tissue Location	Widely expressed. Highest expression is observed in liver and skeletal muscle

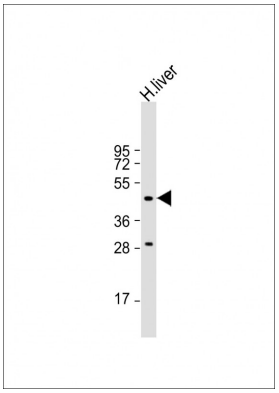
Background

The protein encoded by this gene is involved in the translocation of cobalamin into the mitochondrion, where it is used in the final steps of adenosylcobalamin synthesis. Adenosylcobalamin is a coenzyme required for the activity of methylmalonyl-CoA mutase. Defects in this gene are a cause of methylmalonic aciduria.

References

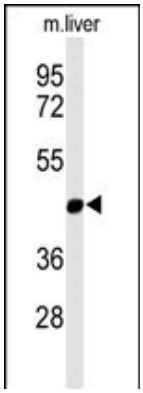
Honjo, R.S., et al. Genet Test Mol Biomarkers 13(2):181-183(2009)
Merinero, B., et al. J. Inherit. Metab. Dis. 31(1):55-66(2008)
Horster, F., et al. Pediatr. Res. 62(2):225-230(2007)
Padovani, D., et al. J. Biol. Chem. 281(26):17838-17844(2006)
Lerner-Ellis, J.P., et al. Hum. Mutat. 24(6):509-516(2004)

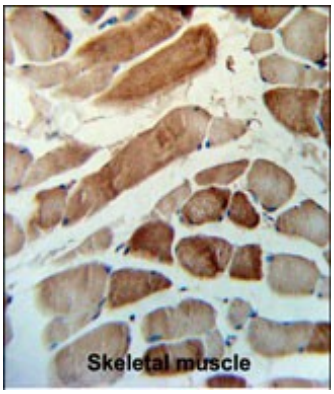
Images



Anti-MMAA Antibody (N-term) at 1:500 dilution + human liver lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 47 kDa Blocking/Dilution buffer: 5% NFDM/TBST.

Western blot analysis of MMAA Antibody (N-term) (Cat. #AP9795a) in mouse liver tissue lysates (35ug/lane). MMAA (arrow) was detected using the purified Pab.





MMAA Antibody (N-term) (Cat. #AP9795a) IHC analysis in formalin fixed and paraffin embedded human skeletal muscle tissue followed by peroxidase conjugation of the secondary antibody and DAB staining. This data demonstrates the use of the MMAA Antibody (N-term) for immunohistochemistry. Clinical relevance has not been evaluated.