

AMACR Recombinant Rabbit mAb

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Catalog # AP94317

Product Information

Application	WB, IHC-P, IHC-F, IF
Reactivity	Human
Host	Rabbit
Clonality	Recombinant
Physical State	Liquid
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Peroxisome. Mitochondrion.
SIMILARITY	Belongs to the CaiB/BaiF CoA-transferase family.
DISEASE	Alpha-methylacyl-CoA racemase deficiency (AMACRD) [MIM:614307]: A rare autosomal recessive peroxisomal disorder characterized by elevated plasma concentrations of pristanic acid C27-bile-acid intermediates, and adult onset of variable neurodegenerative symptoms affecting the central and peripheral nervous systems. Features may include seizures, visual failure, sensorimotor neuropathy, spasticity, migraine, and white matter hyperintensities on brain imaging. Note=The disease is caused by mutations affecting the gene represented in this entry. Congenital bile acid synthesis defect 4 (CBAS4) [MIM:214950]: A disorder characterized by the presence of trihydroxycoprostanic acid in the bile and absence of cholic acid. Patients manifest neonatal jaundice, intrahepatic cholestasis and bile duct deficiency. Note=The disease is caused by mutations affecting the gene represented in this entry.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes a racemase. The encoded enzyme interconverts pristanoyl-CoA and C27-bile acylCoAs between their (R)- and (S)-stereoisomers. The conversion to the (S)-stereoisomers is necessary for degradation of these substrates by peroxisomal beta-oxidation. Encoded proteins from this locus localize to both mitochondria and peroxisomes. Mutations in this gene may be associated with adult-onset sensorimotor neuropathy, pigmentary retinopathy, and adrenomyeloneuropathy due to defects in bile acid synthesis. Alternatively spliced transcript variants have been described. Read-through transcription also exists between this gene and the upstream neighboring C1QTNF3 (C1q and tumor necrosis factor related protein 3) gene. [provided by RefSeq, Mar 2011]

Additional Information

Dilution WB=1:500-1:1000,IHC-P=1:100-500,IHC-F=1:100-500,IF=0

Format

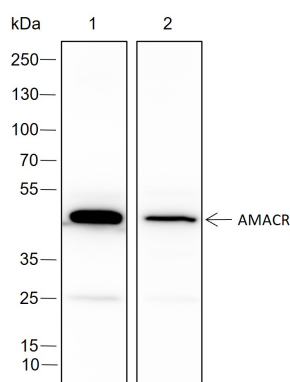
0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glycerol

Storage

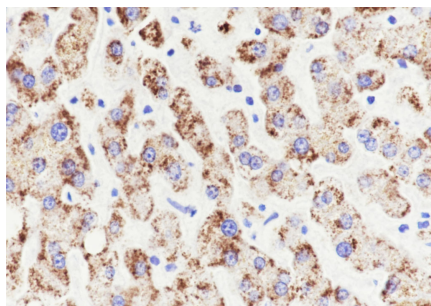
Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Background

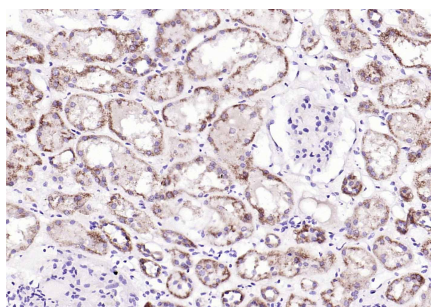
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Images

Blocking buffer: 5% NFDM/TBST Primary ab dilution: 1:1000 Primary ab incubation condition: 2 hours at room temperature Secondary ab: Goat Anti-Rabbit IgG H&L (HRP) Lysate: 1: LNCAP, 2: Human liver Protein loading quantity: 20 µg Exposure time: 30s Predicted MW: 42 kDa Observed MW: 42 kDa



Tissue: Human liver Section type: Formalin-fixed & Paraffin-embedded section Retrieval method: High temperature and high pressure Retrieval buffer: Tris/EDTA buffer, pH 9.0 Primary Ab dilution: 1:100 Primary Ab incubation condition: 1 hour at room temperature Secondary Ab: SP Kit(Rabbit) (sp-0023) Counter stain: Hematoxylin (Blue) Comment: Color brown is the positive signal for AP94317



Paraformaldehyde-fixed, paraffin embedded (human kidney); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Incubation with (AMACR) Monoclonal Antibody, Unconjugated (AP94317) at 1:100 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.