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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.
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

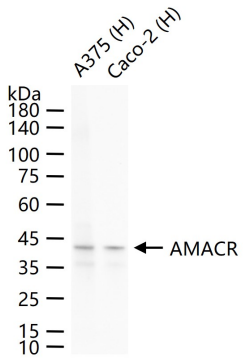
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When

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.

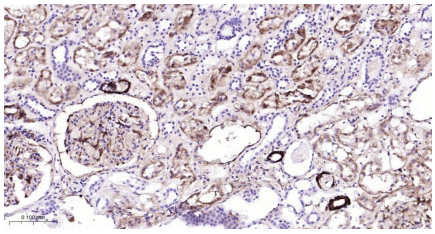
Background

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Images



25 ug total protein per lane of various lysates (see on figure) probed with AMACR monoclonal antibody, unconjugated (AP94317) at 1:1000 dilution and 4°C overnight incubation. Followed by conjugated secondary antibody incubation at r.t. for 60 min.



Paraformaldehyde-fixed, paraffin embedded Human Kidney; Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15 min; Antibody incubation with AMACR Monoclonal Antibody, Unconjugated(AP94317) at 1:100 overnight at 4°C, followed by conjugation to the AP94317-HRP and DAB (C-0010) staining.

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