

AMACR(C-term) mouse mAb

Catalog No: YM1287

Reactivity: Rat

Applications: WB;ICC

Target: AMACR

Fields: >>Primary bile acid biosynthesis;>>Metabolic pathways;>>Peroxisome

Gene Name: amacr

Human Gene Id: 23600

Human Swiss Prot

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No:

Mouse Swiss Prot

No:

Immunogen: Purified recombinant human AMACR(C-terminus) protein fragments expressed

in E.coli.

Q9UHK6

O09174

Specificity: This antibody detects endogenous levels of AMACR(C-terminus) and does not

cross-react with related proteins.

Formulation: Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Source: Monoclonal, Mouse

Dilution: wb 1:1000 icc 1:100

Purification: The antibody was affinity-purified from mouse ascites by affinity-

chromatography using epitope-specific immunogen.

Concentration: 1 mg/ml

Storage Stability: -15°C to -25°C/1 year(Do not lower than -25°C)

Observed Band: 42kD

1/3

Cell Pathway: Primary bile acid biosynthesis;

Background: 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],

Function: catalytic activity:(2S)-2-methylacyl-CoA = (2R)-2-methylacyl-

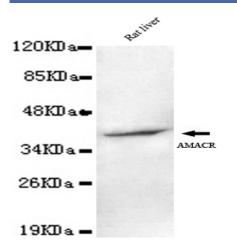
CoA., disease: Defects in AMACR are the cause of alpha-methylacyl-CoA racemase deficiency (AMACRD) [MIM:604489]. AMACRD results in elevated plasma concentrations of pristanic acid C27-bile-acid intermediates. It can be associated with polyneuropathy, retinitis pigmentosa, epilepsy., disease: Defects in AMACR are the cause of congenital bile acid synthesis defect type 4 (CBAS4) [MIM:214950]; also known as cholestasis, intrahepatic, with defective conversion of trihydroxycoprostanic acid to cholic acid or trihydroxycoprostanic acid in bile. Clinical features include neonatal jaundice, intrahepatic cholestasis, bile duct deficiency and absence of cholic acid from bile., function: Racemization of 2-methyl-branched fatty acid CoA esters. Responsible for the conversion of pristanoyl-CoA and C27-bile acyl-CoAs to their (S)-stereoisomers., pa

Subcellular Location :

Peroxisome . Mitochondrion .

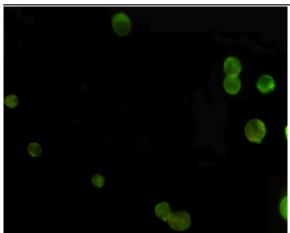
Expression : Aorta, Brain, Cerebellum, Kidney, Liver, PCR rescued clones, Prostate cancer, Sali

Products Images



Western blot detection of AMACR(C-terminus) in Rat Liver lysates using AMACR(C-terminus) mouse mAb (1:1000 diluted). Predicted band size: 42KDa. Observed band size: 42KDa.





Immunocytochemistry staining of Jurkat cells fixed with -20°C Ethanol and using anti-AMACR (C-terminus) mouse mAb (dilution 1:100).