

P504S rabbit pAb

Catalog No :	YT7854
Reactivity :	Human;Mouse;Rat
Applications :	WB;ELISA
Target :	AMACR
Fields :	>>Primary bile acid biosynthesis;>>Metabolic pathways;>>Peroxisome
Gene Name :	AMACR
Protein Name :	P504S
Human Gene Id :	23600
Human Swiss Prot No :	Q9UHK6
Mouse Gene Id :	17117
Mouse Swiss Prot No :	O09174
Rat Swiss Prot No :	P70473
Immunogen :	Synthesized peptide derived from human P504S AA range: 271-320
Specificity :	This antibody detects endogenous levels of Human,Mouse,Rat P504S
Formulation :	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source :	Polyclonal, Rabbit,IgG
Dilution :	WB 1:1000-2000 ELISA 1:5000-20000
Purification :	The antibody was affinity-purified from rabbit antiserum 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)

Molecularweight : 42kD

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 .

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