P504S rabbit pAb

Catalog No	YP-Ab-04360
Isotype	IgG
Reactivity	Human;Mouse;Rat
Applications	WB; ELISA
Gene Name	AMACR
Protein Name	P504S
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
Purification	The antibody was affinity-purified from rabbit serum by affinity-chromatography using specific immunogen.
Dilution	WB 1:1000-2000 ELISA 1:5000-20000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	Alpha-methylacyl-CoA racemase (EC 5.1.99.4;2-methylacyl-CoA racemase)
Observed Band	
Cell Pathway	Peroxisome . Mitochondrion .
Tissue Specificity	
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
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],
matters needing attention	Avoid repeated freezing and thawing!
Usage suggestions	This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

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