## Immunotag<sup>™</sup> P504S Monoclonal Antibody

Antibody Specification	
Catalog No.	ITM1068
Product Description	Immunotag™ P504S Monoclonal Antibody
Size	50 μg, 100 μg
Conjugation	HRP, Biotin, FITC, Alexa Fluor® 350, Alexa Fluor® 405, Alexa Fluor® 488, Alexa Fluor® 555, Alexa Fluor® 594, Alexa Fluor® 647
IMPORTANT NOTE	This product is custom manufactured with a lead time of 3-4 weeks. Once in production, this item cannot be cancelled from an order and is not eligible for return.
Target Protein	P504S
Clonality	Monoclonal
Storage/Stability	-20°C/1 year
Application	WB,IF
Recommended Dilution	Western Blot: 1/1000 - 1/2000. Immunofluorescence: 1/100 - 1/500. Not yet tested in other applications.
Concentration	1 mg/ml
Reactive Species	Human,Mouse,Rat
Host Species	Mouse
Immunogen	Purified recombinant human P504S (C-terminus) protein fragments expressed in Ecoli
Specificity	P504S Monoclonal Antibody detects endogenous levels of P504S protein.
Purification	Affinity purification
Form	Purified mouse monoclonal in buffer containing 0.1M Tris-Glycine (pH 7.4, 150 mM NaCl) with 0.2% sodium azide, 50% glycerol.
Gene Name	AMACR
Accession No.	Q9UHK6 O09174 P70473
Alternate Names	AMACR; Alpha-methylacyl-CoA racemase; 2-methylacyl-CoA racemase

Antibody Specification	
Description	alpha-methylacyl-CoA racemase(AMACR) Homo sapiens 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],
Cell Pathway/ Category	Primary bile acid biosynthesis,
Protein Expression	Aorta,Brain,Cerebellum,Kidney,Liver,PCR rescued clones,Prostate cancer,Sali
Subcellular Localization	cytoplasm,mitochondrion,peroxisome,peroxisomal matrix,
Protein 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., pathway:Lipid metabolism; bile acid biosynthesis., pathway:Lipid metabolism; fatty acid metabolism., similarity:Belongs to the caiB/baiF CoA-transferase family., similarity:Contains 1 C1q domain., similarity:Contains 1 collagen-like domain.,
Usage	For Research Use Only! Not for diagnostic or therapeutic procedures.

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