

IVD Rabbit pAb

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Catalog # AP56597

Product Information

Application	WB
Primary Accession	P26440
Reactivity	Mouse
Predicted	Rat, Pig, Dog, Human, Rabbit, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	46651
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human IVD
Epitope Specificity	201-300/423
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	Mitochondrion matrix.
DISEASE	Defects in IVD are the cause of isovaleric acidemia (IVA) [MIM:243500]. IVA is characterized by retarded psychomotor development, a peculiar odor resembling sweaty feet, an aversion to dietary protein, and pernicious vomiting, leading to acidosis and coma. The acute neonatal form leads to massive metabolic acidosis from the first days of life and rapid death.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Isovaleryl-CoA dehydrogenase (IVD) is a mitochondrial matrix enzyme that catalyzes the third step in leucine catabolism. The genetic deficiency of IVD results in an accumulation of isovaleric acid, which is toxic to the central nervous system and leads to isovaleric acidemia. Alternatively spliced transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Apr 2009]

Additional Information

Gene ID	3712
Other Names	Isovaleryl-CoA dehydrogenase, mitochondrial, IVD, 1.3.8.4, Butyryl-CoA dehydrogenase, 1.3.8.1, IVD (HGNC:6186)
Dilution	WB=1:500-2000
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.

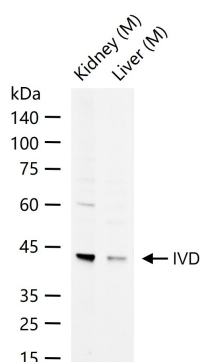
Protein Information

Name	IVD (HGNC:6186)
Function	A mitochondrial matrix enzyme that catalyzes the third step in leucine catabolism, where isovaleryl-CoA (3-methylbutanoyl-CoA) is metabolized to 3-methylbut-2-enoyl-CoA (PubMed: 7640268). To a lesser extent, it also participates in the first step in fatty acid beta- oxidation, in which it catalyzes the proR-proR stereospecific alpha,beta-dehydrogenation of other saturated short-chain acyl-CoA thioesters such as pentanoyl-CoA, hexanoyl-CoA and butanoyl-CoA, using the electron transfer flavoprotein (ETF) as their physiologic electron acceptor(PubMed: 7640268).
Cellular Location	Mitochondrion matrix {ECO:0000250 UniProtKB:P12007}

Background

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Images



25 ug total protein per lane of various lysates (see on figure) probed with IVD polyclonal antibody, unconjugated (AP56597) at 1:1000 dilution and 4°C overnight incubation. Followed by conjugated secondary antibody incubation at r.t. for 60 min.

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