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HIBADH Rabbit pAb

Catalog # AP96925

Product Information

Application	WB
Primary Accession	P31937
Other Accession	P31937
Reactivity	Human, Mouse
Predicted	Rat, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	35329
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human HIBADH
Epitope Specificity	251-336/336
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the 3-hydroxyisobutyrate dehydrogenase family.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	HIBADH is a 336 amino acid mitochondrial enzyme that catalyzes the NAD+-dependent, reversible oxidization of 3-Hydroxyisobutyrate to methylmalonate semialdehyde, an intermediate of valine catabolism. The enzyme functions as a homodimer between a pH of 7.0 and 10.0, with optimal activity between 8.8 and 9.0. It was previously hypothesized that defects in the gene encoding HIBADH may be the cause of 3-Hydroxyisobutyric aciduria, a rare disorder that is characterized by a variety of clinical manifestations such as neurodevelopmental problems and dysmorphic features. However, it was shown that HIBADH activity was equal in patients with 3-Hydroxyisobutyric aciduria as compared with controls.

Additional Information

Gene ID	11112
Other Names	3HIDH_HUMAN; HIBADH; 1.1.1.31; 3HIDH_MOUSE; 3HIDH_RAT; 3-hydroxyisobutyrate dehydrogenase
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	HIBADH
Cellular Location	Mitochondrion.
Tissue Location	Detected in skin fibroblasts.

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