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L2HGDH Rabbit pAb

Catalog # AP98119

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q9H9P8
Other Accession	Q9H9P8
Reactivity	Mouse
Predicted	Rat, Pig, Orangutan, Human, Rabbit, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	50316
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human L2HGDH
Epitope Specificity	201-300/463
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 L2HGDH family.
DISEASE	Defects in L2HGDH are the cause of L-2-hydroxyglutaric aciduria (L2HGA) [MIM:236792]. L2HGA is a rare autosomal recessive disorder clinically characterized by mild psychomotor delay in the first years of life, followed by progressive cerebellar ataxia, dysarthria and moderate to severe mental retardation. Diagnosis is based on the presence of an excess of L-2-hydroxyglutaric acid in urine, blood and cerebrospinal fluid.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes L-2-hydroxyglutarate dehydrogenase, a FAD-dependent enzyme that oxidizes L-2-hydroxyglutarate to alpha-ketoglutarate in a variety of mammalian tissues. Mutations in this gene cause L-2-hydroxyglutaric aciduria, a rare autosomal recessive neurometabolic disorder resulting in moderate to severe mental retardation. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	79944
Other Names	C14orf160; L2HGA; RGD1306250; L2HDH_BOVIN; L2HGDH; Duranin; 1.1.99.2; L2HDH_HUMAN; L2HDH_MOUSE; L2HDH_PONAB; L-2-hydroxyglutarate dehydrogenase; chromosome 14 open reading frame 160; 2-hydroxyglutarate dehydrogenase
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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	L2HGDH (HGNC:20499)
Function	Catalyzes the oxidation of L-2-hydroxyglutarate to 2- oxoglutarate (alpha-ketoglutarate).
Cellular Location	Mitochondrion.
Tissue Location	Widely expressed. Highly expressed in brain, testis and muscle. Expressed to a lower extent in lymphocytes, fibroblasts, keratinocytes, placenta, bladder, small intestine, liver and bone marrow.

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