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Rabbit Anti-L2HGDH antibody

SL16876R

Product Name:	L2HGDH
Chinese Name:	L2HGDH蛋白抗体
Alias:	2 hydroxyglutarate dehydrogenase; Alpha hydroxyglutarate oxidoreductase; Alpha ketoglutarate reductase; C14orf160; Duranin; FLJ12618; L alpha hydroxyglutarate dehydrogenase; L-2-hydroxyglutarate dehydrogenase; L-2-hydroxyglutarate dehydrogenase, mitochondrial; L2HDH_HUMAN; l2hgdh; mitochondrial.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Pig,Cow,Horse,Rabbit,Sheep,Orangutan
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	45kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human L2HGDH:201-300/463
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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]

Subcellular Location:
Mitochondrion.

Tissue Specificity:
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.

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.

Similarity:
Belongs to the L2HGDH family.

SWISS:
Q9H9P8

Gene ID:
79944

Database links:

Entrez Gene: 514230 Cow

Entrez Gene: 79944 Human

Entrez Gene: 217666 Mouse

Entrez Gene: 100152180 Pig

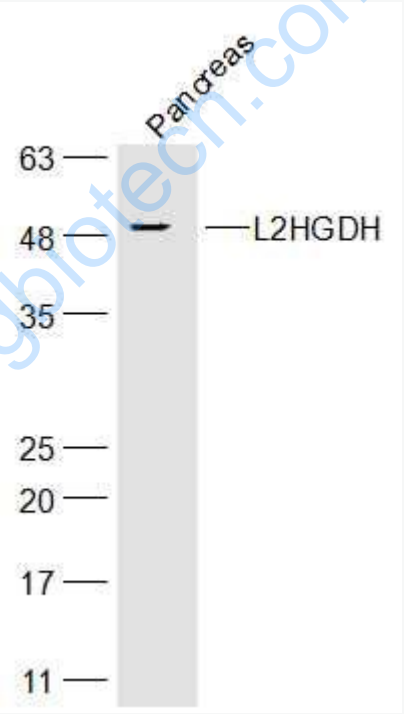
Entrez Gene: 314196 Rat

Omim: 609584 Human

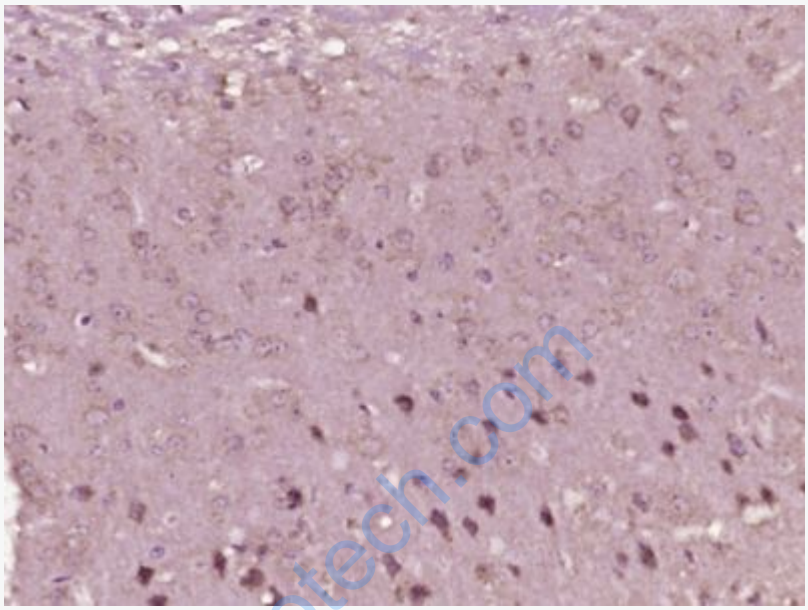
SwissProt: A7MBI3 Cow

SwissProt: Q9H9P8 Human

SwissProt: Q91YP0 Mouse

	SwissProt: Q5R9N7 Orangutan Unigene: 256034 Human Unigene: 103362 Mouse Unigene: 22733 Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Sample: Pancreas (Mouse) Lysate at 40 ug Primary: Anti-L2HGDH (SL16876R) at 1/300 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 45 kD

Observed band size: 48 kD



Paraformaldehyde-fixed, paraffin embedded (Mouse brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (L2HGDH) Polyclonal Antibody, Unconjugated (SL16876R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.