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Rabbit Anti-GCDH antibody

SL13312R

Product Name:	GCDH
Chinese Name:	戊二酰辅酶A脱氢酶抗体
Alias:	ACAD5; EC 1.3.99.7; GCD; Gcdh; GCDH_HUMAN; Glutaryl CoA dehydrogenase, mitochondrial; Glutaryl Coenzyme A dehydrogenase; Glutaryl-CoA dehydrogenase; mitochondrial; MS781.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,
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:	43kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human GCDH:201-300/438
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:	GCDH is a 438 amino acid protein that localizes to the mitochondrial matrix and belongs to the acyl-CoA dehydrogenase family. Existing as a homotetramer, GCDH uses FAD as a cofactor to catalyze the oxidative decarboxylation of glutaryl-CoA to crotonyl-CoA and CO(2) in the degradative pathway of L-lysine, L-hydroxylysine and L-tryptophan metabolism. While GCDH exists as both a long and short isoform, only

	the long isoform is a functionally active protein. Defects in the gene encoding GCDH are the cause of glutaric acidemia type I (GA-I), an autosomal recessive disorder that is characterized by the accumulation of glutaconic acid and is associated with such symptoms as progressive dystonia and athetosis due to gliosis and neuronal loss in the basal ganglia. **Function:** Catalyzes the oxidative decarboxylation of glutaryl-CoA to crotonyl-CoA and CO(2) in the degradative pathway of L-lysine, L-hydroxylysine, and L-tryptophan metabolism. It uses electron transfer flavoprotein as its electron acceptor. Isoform Short is inactive. **Subunit:** Homotetramer. **Subcellular Location:** Mitochondrion matrix. **Tissue Specificity:** Isoform 1 and isoform 2 are expressed in fibroblasts and liver. **DISEASE:** Defects in GCDH are the cause of glutaric aciduria type 1 (GA1) [MIM:231670]. GA1 is an autosomal recessive metabolic disorder characterized by progressive dystonia and athetosis due to gliosis and neuronal loss in the basal ganglia. **Similarity:** Belongs to the acyl-CoA dehydrogenase family. **SWISS:** Q92947 **Gene ID:** 2639 **Database links:** Entrez Gene: 2639Human Entrez Gene: 270076Mouse Entrez Gene: 364975Rat Omim: 608801Human SwissProt: Q92947Human SwissProt: Q60759Mouse

	Important Note: This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.