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Rabbit Anti-GCSH antibody

SL13323R

Product Name:	GCSH
Chinese Name:	甘氨酸裂解系统H蛋白抗体
Alias:	GCE; GCSH; GCSH_HUMAN; Glycine cleavage system H protein; Glycine cleavage system H protein mitochondrial; Glycine cleavage system protein H (aminomethyl carrier); Glycine cleavage system protein H; Lipoic acid containing protein; mitochondrial; Mitochondrial glycine cleavage system H protein; NKH.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Horse,
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:	14kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human GCSH:101-173/173
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:	GCSH is a 173 amino acid mitochondrial protein that contains one lipoyl-binding domain and belongs to the gcvH family. Defects in the gene encoding GCSH are the cause of glycine encephalopathy (GCE), an autosomal recessive disease that is also referred to as non-ketotic hyperglycinemia (NKH). Characterized by severe

neurological symptoms, patients with GCE have a large amount of glycine accumulated in their body fluids. The gene encoding GCSH maps to human chromosome 16, which encodes over 900 genes and comprises nearly 3% of the human genome.

Function:
The glycine cleavage system catalyzes the degradation of glycine. The H protein shuttles the methylamine group of glycine from the P protein to the T protein.

Subcellular Location:
Mitochondrion.

DISEASE:
Defects in GCSH are a cause of non-ketotic hyperglycinemia (NKH) [MIM:605899]; also known as glycine encephalopathy (GCE). NKH is an autosomal recessive disease characterized by accumulation of a large amount of glycine in body fluid and by severe neurological symptoms.

Similarity:
Belongs to the gcvH family.
Contains 1 lipoyl-binding domain.

SWISS:
P23434

Gene ID:
2653

Database links:

Entrez Gene: 317723Cow

Entrez Gene: 2653Human

Entrez Gene: 68133Mouse

Entrez Gene: 171133Rat

Omim: 238330Human

SwissProt: P20821Cow

SwissProt: P23434Human

SwissProt: Q91WK5Mouse

SwissProt: Q5I0P2Rat

Important Note:

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