antibodies .- online.com





anti-GCSH antibody (AA 101-173) (Alexa Fluor 750)



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| Quantity: | 100 μL | |
|----------------------|--|--|
| Target: | GCSH | |
| Binding Specificity: | AA 101-173 | |
| Reactivity: | Human | |
| Host: | Rabbit | |
| Clonality: | Polyclonal | |
| Conjugate: | This GCSH antibody is conjugated to Alexa Fluor 750 | |
| Application: | Western Blotting (WB), Immunofluorescence (Cultured Cells) (IF (cc)), Immunofluorescence (Paraffin-embedded Sections) (IF (p)) | |

Product Details

| Immunogen: | KLH conjugated synthetic peptide derived from human GCSH |
|-----------------------|--|
| Isotype: | IgG |
| Predicted Reactivity: | Human,Mouse,Rat,Horse |
| Purification: | Purified by Protein A. |

Target Details

| Target: | GCSH |
|-------------------|--|
| Alternative Name: | GCSH (GCSH Products) |
| Background: | Synonyms: 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.

Background: 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.

Gene ID:

2653

Application Details

Application Notes: IF(IHC-P) 1:50-200

IF(IHC-F) 1:50-200 IF(ICC) 1:50-200

Restrictions:

For Research Use only

Handling

| Format: | Liquid |
|--------------------|--|
| Concentration: | 1 μg/μL |
| Buffer: | Aqueous buffered solution containing 0.01M TBS (pH 7.4) with 1 % BSA, 0.03 % Proclin300 and 50 % Glycerol. |
| Preservative: | ProClin |
| Precaution of Use: | This product contains ProClin: a POISONOUS AND HAZARDOUS SUBSTANCE, which should be handled by trained staff only. |
| Storage: | -20 °C |
| Storage Comment: | Store at -20°C. Aliquot into multiple vials to avoid repeated freeze-thaw cycles. |
| Expiry Date: | 12 months |