bs-13323R **[Primary Antibody]**

GCSH Rabbit pAb

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DATASHEET

Host: Rabbit **Isotype:** IgG

Clonality: Polyclonal

GeneID: 2653 **SWISS:** P23434

Target: GCSH

Immunogen: KLH conjugated synthetic peptide derived from human GCSH: 101-173/173.

Purification: affinity purified by Protein A

Concentration: 1mg/ml

Storage: 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
Shipped at 4°C. Store at -20°C for one year. Avoid repeated freeze/thaw cycles.

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.

Applications: **WB** (1:500-2000)
ELISA (1:5000-10000)

Reactivity: (predicted: Human, Mouse, Rat, Horse)

Predicted MW.: 14 kDa

Subcellular Location: Cytoplasm