bs-13322R **[Primary Antibody]**

GCS1 Rabbit pAb

www.bioss.com.cn
sales@bioss.com.cn
techsupport@bioss.com.cn
400-901-9800

DATASHEET

Host: Rabbit **Isotype:** IgG

Clonality: Polyclonal

GeneID: 7841 **SWISS:** Q13724

Target: GCS1

Immunogen: KLH conjugated synthetic peptide derived from human GCS1: 51-150/837.

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: Glycosylation of asparagine residues in Asn-X-Ser/Thr motifs in proteins commonly occur in the lumen of the endoplasmic reticulum (ER). Glucosidase I catalyzes the first step in the N-linked oligosaccharide processing pathway. It specifically removes the distal alpha 1,2-linked glucose residue from the Glc3-Man9-GlcNAc2 oligosaccharide precursor. Glucosidase I contains a short cytosolic tail, a single pass transmembrane domain and a large C-terminal catalytic domain located on the luminal side of the ER. Mutations in the gene encoding Glucosidase I result in the congenital disorder glycosylation (CDG-IIb), which is characterized by generalized hypotonia, dysmorphic features, hepatomegaly, hypoventilation, feeding problems, seizures and death. Two point mutations in the Glucosidase I gene have been identified and result in amino acid substitutions, namely Arg486Thr and Phe652Leu, that affect polypeptide folding and active site formation.

Applications: **WB** (1:500-2000)
IHC-P (1:100-500)
IHC-F (1:100-500)
IF (1:100-500)
ICC/IF (1:100-500)
ELISA (1:5000-10000)

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

Predicted MW.: 92 kDa

Subcellular Location: Cell membrane ,Cytoplasm