


## Product datasheet

# Anti-GCS1 antibody ab82962

1 Image

### Overview

<b>Product name</b>	Anti-GCS1 antibody
<b>Description</b>	Rabbit polyclonal to GCS1
<b>Tested applications</b>	<b>Suitable for:</b> WB, ELISA
<b>Species reactivity</b>	<b>Reacts with:</b> Human <b>Predicted to work with:</b> Mouse, Rat, Rabbit, Horse, Guinea pig, Cow, Cat, Dog, Zebrafish 
<b>Immunogen</b>	Synthetic peptide corresponding to a region within N terminal amino acids 144-193 (GPYGWEFHGDG LSFGRQHIQD GALRLTTEFV KRPGGQHGGD WSWRVTVEPQ) of human GCS1 (NP_006293). <a href="#">Run BLAST with ExPASy</a> <a href="#">Run BLAST with NCBI</a>
<b>Positive control</b>	HeLa cell lysate.

### Properties

<b>Form</b>	Liquid
<b>Storage instructions</b>	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid repeated freeze / thaw cycles.
<b>Storage buffer</b>	Preservative: None Constituents: 2% Sucrose, PBS
<b>Purity</b>	Immunogen affinity purified
<b>Purification notes</b>	ab82962 is purified by a peptide affinity chromatography method.
<b>Clonality</b>	Polyclonal
<b>Isotype</b>	IgG

### Applications

Our [Abpromise guarantee](#) covers the use of **ab82962** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

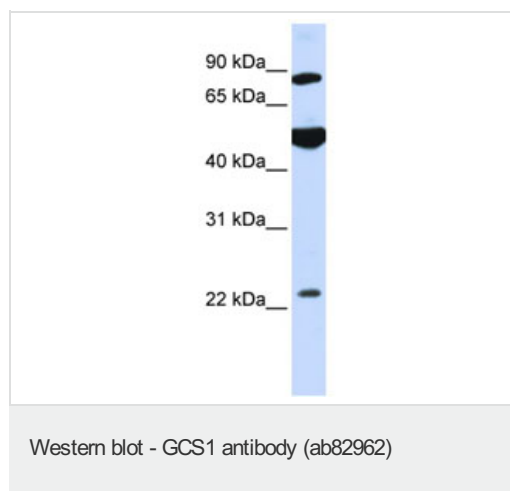
Application	Abreviews	Notes

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WB		Use a concentration of 1 µg/ml. Predicted molecular weight: 92 kDa. Good results were obtained when blocked with 5% non-fat dry milk in 0.05% PBS-T.
ELISA		Use at an assay dependent concentration. ELISA titre using peptide based assay, 1:312500.

## Target

<b>Function</b>	Cleaves the distal alpha 1,2-linked glucose residue from the Glc(3)Man(9)GlcNAc(2) oligosaccharide precursor in a highly specific manner.
<b>Pathway</b>	Glycan metabolism; N-glycan degradation.
<b>Involvement in disease</b>	Defects in MOGS are the cause of type IIb congenital disorder of glycosylation (CDGIIb) [MIM:606056]; also known as glucosidase I deficiency. CDGIIb is characterized by marked generalized hypotonia and hypomotility of the neonate, dysmorphic features, including a prominent occiput, short palpebral fissures, retrognathia, high arched palate, generalized edema, and hypoplastic genitalia. Symptoms of the infant included hepatomegaly, hypoventilation, feeding problems and seizures. The clinical course was progressive and the infant did not survive more than a few months.
<b>Sequence similarities</b>	Belongs to the glycosyl hydrolase 63 family.
<b>Cellular localization</b>	Endoplasmic reticulum membrane.

## Images



Anti-GCS1 antibody (ab82962) at 1 µg/ml +  
HeLa cell lysate at 10 µg

### Secondary

HRP conjugated anti-Rabbit IgG at 1/50000  
dilution

**Predicted band size** : 92 kDa

**Observed band size** : 92 kDa

**Additional bands at** : 24 kDa, 50 kDa. We  
are unsure as to the identity of these extra  
bands.

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