




## Product datasheet

### Anti-GNS antibody ab230044

3 Images

#### Overview

<b>Product name</b>	Anti-GNS antibody
<b>Description</b>	Rabbit polyclonal to GNS
<b>Host species</b>	Rabbit
<b>Tested applications</b>	<b>Suitable for:</b> WB, IHC-P
<b>Species reactivity</b>	<b>Reacts with:</b> Mouse, Human <b>Predicted to work with:</b> Goat, Cow 
<b>Immunogen</b>	Recombinant fragment corresponding to Human GNS aa 250 to the C-terminus. Database link: <a href="#">P15586</a>  <a href="#">Run BLAST with</a>  <a href="#">Run BLAST with</a>
<b>Positive control</b>	WB: Mouse liver tissue lysate. IHC-P: Human testis and kidney tissues.
<b>General notes</b>	<p>The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing.</p> <p>If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&amp;As</p>

#### Properties

<b>Form</b>	Liquid
<b>Storage instructions</b>	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle.
<b>Storage buffer</b>	pH: 7.30 Preservative: 0.02% Sodium azide Constituents: PBS, 50% Glycerol (glycerin, glycerine)
<b>Purity</b>	Immunogen affinity purified
<b>Clonality</b>	Polyclonal
<b>Isotype</b>	IgG

## Applications

### The Abpromise guarantee

Our **Abpromise guarantee** covers the use of ab230044 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
WB		1/1000 - 1/5000. Predicted molecular weight: 63,60 kDa.
IHC-P		1/20 - 1/200.

## Target

### Involvement in disease

Defects in GNS are the cause of mucopolysaccharidosis type 3D (MPS3D) [MIM:252940]; also known as Sanfilippo D syndrome. MPS3D is a form of mucopolysaccharidosis type 3, an autosomal recessive lysosomal storage disease due to impaired degradation of heparan sulfate. MPS3 is characterized by severe central nervous system degeneration, but only mild somatic disease. Onset of clinical features usually occurs between 2 and 6 years; severe neurologic degeneration occurs in most patients between 6 and 10 years of age, and death occurs typically during the second or third decade of life.

### Sequence similarities

Belongs to the sulfatase family.

### Post-translational modifications

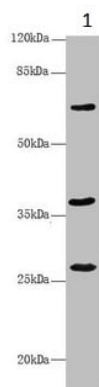
The form A (78 kDa) is processed by internal peptidase cleavage to a 32 kDa N-terminal species (form B) and a 48 kDa C-terminal species.

The conversion to 3-oxoalanine (also known as C-formylglycine, FGly), of a serine or cysteine residue in prokaryotes and of a cysteine residue in eukaryotes, is critical for catalytic activity.

### Cellular localization

Lysosome.

## Images



Western blot - Anti-GNS antibody (ab230044)

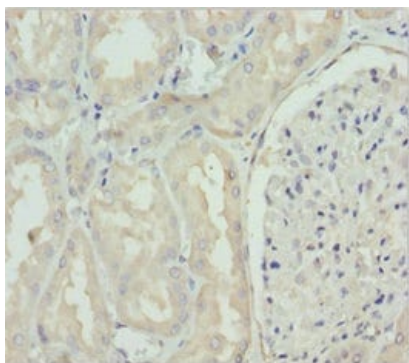
Anti-GNS antibody (ab230044) at 1/1000 dilution + Mouse liver tissue lysate

### Secondary

Goat polyclonal to rabbit IgG at 1/10000 dilution

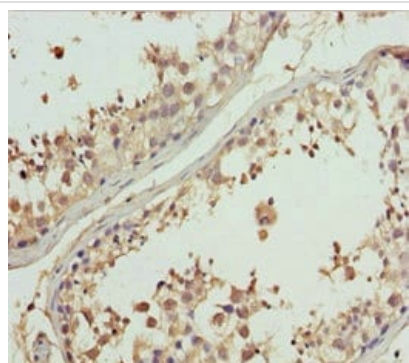
**Predicted band size:** 63,60 kDa

**Observed band size:** 62,27,37 kDa



Paraffin-embedded human kidney tissue stained for GNS using ab230044 at 1/100 dilution in immunohistochemical analysis.

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GNS antibody (ab230044)



Paraffin-embedded human testis tissue stained for GNS using ab230044 at 1/100 dilution in immunohistochemical analysis.

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GNS antibody (ab230044)

**Please note:** All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

### Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit <https://www.abcam.com/abpromise> or contact our technical team.

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