

#### DESCRIPTION

|                           |                                                                                                                                                                                                                                                                             |
|---------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>Species Reactivity</b> | Human                                                                                                                                                                                                                                                                       |
| <b>Specificity</b>        | Detects human $\alpha$ -N-acetylgalactosaminidase/NAGA in direct ELISAs and Western blots. In direct ELISAs, less than 2% cross-reactivity with recombinant human $\alpha$ -Galactosidase A is observed.                                                                    |
| <b>Source</b>             | Polyclonal Sheep IgG                                                                                                                                                                                                                                                        |
| <b>Purification</b>       | Antigen Affinity-purified                                                                                                                                                                                                                                                   |
| <b>Immunogen</b>          | Chinese hamster ovary cell line recombinant human $\alpha$ -N-acetylgalactosaminidase/NAGA<br>Leu18-Gln411<br>Accession # P17050                                                                                                                                            |
| <b>Conjugate</b>          | Alexa Fluor 594<br>Excitation Wavelength: 590 nm<br>Emission Wavelength: 617 nm                                                                                                                                                                                             |
| <b>Formulation</b>        | Supplied 0.2mg/ml in 1X PBS with RDF1 and 0.09% Sodium Azide<br><br>*Contains <0.1% Sodium Azide, which is not hazardous at this concentration according to GHS classifications. Refer to the Safety Data Sheet (SDS) for additional information and handling instructions. |

#### APPLICATIONS

**Please Note:** Optimal dilutions should be determined by each laboratory for each application. [General Protocols](#) are available in the Technical Information section on our website.

**Western Blot** Optimal dilution of this antibody should be experimentally determined.

#### PREPARATION AND STORAGE

**Shipping** The product is shipped with polar packs. Upon receipt, store it immediately at the temperature recommended below.

**Stability & Storage** Protect from light. Do not freeze. 12 months from date of receipt, 2 to 8 °C as supplied

#### BACKGROUND

NAGA is a lysosomal  $\alpha$ -N-acetylgalactosaminidase that cleaves non-reducing  $\alpha$ -N-acetylgalactosaminyl moieties from glycoconjugates (1). Mature NAGA has 394 amino acids and is trafficked to the lysosome via the mannose-6-phosphate receptor-mediated pathway (2). The enzyme is a retaining exoglycosidase, where both the substrate and product of the enzymatic reaction have the same anomeric configuration (3). Deficiency in NAGA results in increased urinary excretion and tissue accumulation of glycopeptides and oligosaccharides containing terminal  $\alpha$ -N-acetylgalactosaminyl moieties (4), manifesting as Schindler's disease, an autosomal recessive disease with neuroaxonal dystrophy and other neurological symptoms (5). The enzyme can be used to remove  $\alpha$ -N-acetylgalactosaminyl residues present on red blood cells thus converting blood type A to blood type O (6, 7, 8).

#### PRODUCT SPECIFIC NOTICES

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