

## DESCRIPTION

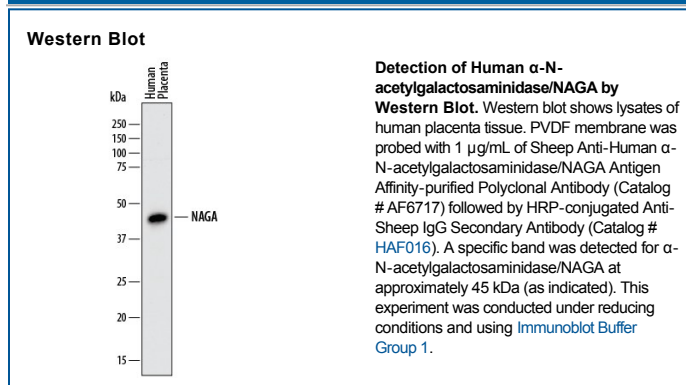
<b>Species Reactivity</b>	Human
<b>Specificity</b>	Detects human $\alpha$ -N-acetylgalactosaminidase/NAGA in direct ELISAs and Western blots. In direct ELISAs, less than 2% cross-reactivity with recombinant human $\alpha$ -Galactosidase A is observed.
<b>Source</b>	Polyclonal Sheep IgG
<b>Purification</b>	Antigen Affinity-purified
<b>Immunogen</b>	Chinese hamster ovary cell line recombinant human $\alpha$ -N-acetylgalactosaminidase/NAGA Leu18-Gln411 Accession # P17050
<b>Formulation</b>	Lyophilized from a 0.2 $\mu$ m filtered solution in PBS with Trehalose. See Certificate of Analysis for details. *Small pack size (-SP) is supplied either lyophilized or as a 0.2 $\mu$ m filtered solution in PBS.

## 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.

	Recommended Concentration	Sample
<b>Western Blot</b>	1 $\mu$ g/mL	See Below

## DATA



## PREPARATION AND STORAGE

<b>Reconstitution</b>	Sterile PBS to a final concentration of 0.2 mg/mL.
<b>Shipping</b>	The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below. *Small pack size (-SP) is shipped with polar packs. Upon receipt, store it immediately at -20 to -70 °C
<b>Stability &amp; Storage</b>	<b>Use a manual defrost freezer and avoid repeated freeze-thaw cycles.</b> <ul style="list-style-type: none"> <li>● 12 months from date of receipt, -20 to -70 °C as supplied.</li> <li>● 1 month, 2 to 8 °C under sterile conditions after reconstitution.</li> <li>● 6 months, -20 to -70 °C under sterile conditions after reconstitution.</li> </ul>

## 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).

### References:

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3. Garman, S.C. *et al.* (2002) Structure. **10**:425.
4. Eng, C.M. *et al.* (2001) N. Engl. J. Med. **345**:9.
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