

DESCRIPTION

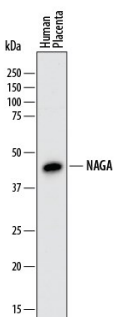
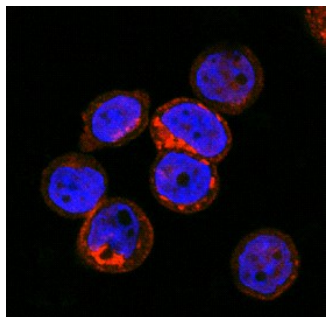
Species Reactivity	Human
Specificity	Detects human α -N-acetylgalactosaminidase/NAGA in direct ELISAs and Western blots. In direct ELISAs, less than 2% cross-reactivity with recombinant human α -Galactosidase A is observed.
Source	Polyclonal Sheep IgG
Purification	Antigen Affinity-purified
Immunogen	Chinese hamster ovary cell line recombinant human α -N-acetylgalactosaminidase/NAGA Leu18-Gln411 Accession # P17050
Formulation	Lyophilized from a 0.2 μ m filtered solution in PBS with Trehalose. See Certificate of Analysis for details. *Small pack size (-SP) is supplied as a 0.2 μ 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
Western Blot	1 μ g/mL	See Below
Immunocytochemistry	5-15 μ g/mL	See Below

DATA

<p>Western Blot</p>  <p>Detection of Human α-N-acetylgalactosaminidase/NAGA by Western Blot. Western blot shows lysates of human placenta tissue. PVDF membrane was probed with 1 μg/mL of Sheep Anti-Human α-N-acetylgalactosaminidase/NAGA Antigen Affinity-purified Polyclonal Antibody (Catalog # AF6717) followed by HRP-conjugated Anti-Sheep IgG Secondary Antibody (Catalog # HAF016). A specific band was detected for α-N-acetylgalactosaminidase/NAGA at approximately 45 kDa (as indicated). This experiment was conducted under reducing conditions and using Immunoblot Buffer Group 1.</p>	<p>Immunocytochemistry</p>  <p>α-N-acetylgalactosaminidase/NAGA in HeLa Human Cell Line. α-N-acetylgalactosaminidase/NAGA was detected in immersion fixed HeLa human cervical epithelial carcinoma cell line using Sheep Anti-Human α-N-acetylgalactosaminidase/NAGA Antigen Affinity-purified Polyclonal Antibody (Catalog # AF6717) at 15 μg/mL for 3 hours at room temperature. Cells were stained using the NorthernLights™ 557-conjugated Anti-Sheep IgG Secondary Antibody (red; Catalog # NL010) and counterstained with DAPI (blue). Specific staining was localized to lysosomes. View our protocol for Fluorescent ICC Staining of Cells on Coverslips.</p>
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PREPARATION AND STORAGE

Reconstitution	Sterile PBS to a final concentration of 0.2 mg/mL.
Shipping	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
Stability & Storage	Use a manual defrost freezer and avoid repeated freeze-thaw cycles. <ul style="list-style-type: none"> ● 12 months from date of receipt, -20 to -70 °C as supplied. ● 1 month, 2 to 8 °C under sterile conditions after reconstitution. ● 6 months, -20 to -70 °C under sterile conditions after reconstitution.

BACKGROUND

NAGA is a lysosomal α -N-acetylgalactosaminidase that cleaves non-reducing α -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 α -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 α -N-acetylgalactosaminyl residues present on red blood cells thus converting blood type A to blood type O (6, 7, 8).

References:

1. Wang, A.M. *et al.* (1990) *J. Biol. Chem.* **265**:21859.
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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.
5. Wang, A.M. *et al.* (1990) *J. Clin. Invest.* **86**:1752.
6. Liu, Q.P. *et al.* (2007) *Nature Biotechnol.* **25**:454.
7. Calcutt, M. J. *et al.* (2002) *FEMS Microbiol. Lett.* **214**:77.
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