

#### DESCRIPTION

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|---------------------------|---|
| <b>Species Reactivity</b> | Human   |
| <b>Specificity</b>        | Detects human Hexosaminidase A/HEXA in direct ELISAs and Western blots.   |
| <b>Source</b>             | Polyclonal Sheep IgG  |
| <b>Purification</b>       | Antigen Affinity-purified   |
| <b>Immunogen</b>          | <i>S. frugiperda</i> insect ovarian cell line Sf 21-derived recombinant human Hexosaminidase A/HEXA<br>Leu23-Thr529<br>Accession # P06865   |
| <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

|                                |   |
|--------------------------------|---|
| <b>Shipping</b>                | The product is shipped with polar packs. Upon receipt, store it immediately at the temperature recommended below. |
| <b>Stability &amp; Storage</b> | Protect from light. Do not freeze. 12 months from date of receipt, 2 to 8 °C as supplied                          |

#### BACKGROUND

HEXA (beta-hexosaminidase subunit alpha; also hexaminidase subunit alpha) is a 50-56 kDa glycoprotein member of the glycosyl hydrolase #20 family of enzymes. It is found in the lysosomes of a variety of cell types including fibroblasts, lacrimal acinar cells, and neurons. β-Hexosaminidase is a multimeric enzyme that cleaves aminoacetylhexosamines from multiple glycosylated molecules. It exists in three noncovalent isozyme forms, among which is an A form that is trimeric and composed of one α- and two β-subunits, a B form that is tetrameric and contains two α- and two β-subunits, and a dimeric S form that contains two α-subunits. The α- and β-subunits are products of related but distinct genes. The β-hexosiminidase A form acts on a wide range of substrates, while the B form activity is biased towards sulfated hexoses. Human HEXA is synthesized as a 529 aa (amino acid) prepro-precursor. It contains a 22 aa signal sequence, a 66 aa prosegment, and a 441 aa mature region that contains a catalytic domain (aa 167-488). The prosegment may undergo additional processing through His90, and this segment (≈ 6 kDa) is known to remain linked to the mature region via a disulfide bond. Notably, circulating HEXA is comprised only of subunit proforms whose MW may run some 10 kDa higher in SDS-PAGE. Over aa 23-529, human HEXA shares 85% and 57% aa sequence identity with mouse HEXA and the human HEXA β-subunit proprecursor, respectively.

#### PRODUCT SPECIFIC NOTICES

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