

#### DESCRIPTION

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|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------------------------------------------------------|
| <b>Species Reactivity</b>                                                                                                                                                                               | Human                                                                                                                                 |
| <b>Specificity</b>                                                                                                                                                                                      | Detects human $\alpha$ -L-Iduronidase/IDUA in direct ELISAs and Western blots.                                                        |
| <b>Source</b>                                                                                                                                                                                           | Polyclonal Sheep IgG                                                                                                                  |
| <b>Purification</b>                                                                                                                                                                                     | Antigen Affinity-purified                                                                                                             |
| <b>Immunogen</b>                                                                                                                                                                                        | Mouse myeloma cell line NS0-derived recombinant human $\alpha$ -L-Iduronidase/IDUA<br>Ala26-Pro653 (Ala26Thr)<br>Accession # AAA81589 |
| <b>Conjugate</b>                                                                                                                                                                                        | Alexa Fluor 647<br>Excitation Wavelength: 650 nm<br>Emission Wavelength: 668 nm                                                       |
| <b>Formulation</b>                                                                                                                                                                                      | Supplied 0.2mg/ml in 1X PBS with RDF1 and 0.09% Sodium Azide                                                                          |
| *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.

|                            |                                                                        |
|----------------------------|------------------------------------------------------------------------|
| <b>Western Blot</b>        | Optimal dilution of this antibody should be experimentally determined. |
| <b>Immunocytochemistry</b> | Optimal dilution of this antibody should be experimentally determined. |
| <b>Immunoprecipitation</b> | 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

$\alpha$ -L-Iduronidase encoded by the IDUA gene is an important enzyme required for the lysosomal degradation of glycosaminoglycans (GAGs). It hydrolyzes the non-reducing terminal  $\alpha$ -L-iduronic acid residues in GAGs including dermatan sulfate and heparan sulfate. Mutations in IDUA that result in enzymatic deficiency lead to the autosomal recessive disease mucopolysaccharidosis type I (MPS I) (1). MPS I causes progressive cellular, tissue and organ damage, and several clinical studies using enzyme replacement therapy have shown promising benefits (2).

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