

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human α -L-Iduronidase/IDUA in direct ELISAs and Western blots.
Source	Polyclonal Sheep IgG
Purification	Antigen Affinity-purified
Immunogen	Mouse myeloma cell line NS0-derived recombinant human α -L-Iduronidase/IDUA Ala26-Pro653 (Ala26Thr) Accession # AAA81589
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.

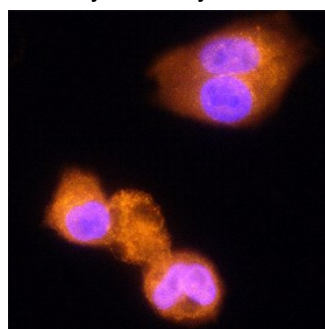
APPLICATIONS

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	0.1 μ g/mL	Recombinant Human α -L-Iduronidase/IDUA (Catalog # 4119-GH)
Immunocytochemistry	5-15 μ g/mL	See Below
Immunoprecipitation	25 μ g/mL	Conditioned cell culture medium spiked with Recombinant Human α -L-Iduronidase/IDUA (Catalog # 4119-GH), see our available Western blot detection antibodies

DATA

Immunocytochemistry



α -L-Iduronidase/IDUA in HepG2 Human Cell Line.

α -L-Iduronidase/IDUA was detected in immersion fixed HepG2 human hepatocellular carcinoma cell line using Sheep Anti-Human α -L-Iduronidase/IDUA Antigen Affinity-purified Polyclonal Antibody (Catalog # AF4119) at 15 μ g/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 cytoplasm. View our protocol for [Fluorescent ICC Staining of Cells on Coverslips](#).

PREPARATION AND STORAGE

Reconstitution	Reconstitute at 0.2 mg/mL in sterile PBS.
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

α -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 α -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).

References:

1. Scott, H.S. *et al.* (1995) Hum. Mutat. **6**:288.
2. Wraith, J.E. (2005) Expert Opin. Pharmacother. **6**:489.