Human α-L-Iduronidase/IDUA Antibody

Monoclonal Mouse IgG_1 Clone # 452619

Catalog Number: MAB4119

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human α-L-Iduronidase/IDUA in direct ELISAs and Western blots.
Source	Monoclonal Mouse IgG_1 Clone # 452619
Purification	Protein A or G purified from hybridoma culture supernatant
Immunogen	Mouse myeloma cell line NS0-derived recombinant human α-L-Iduronidase/IDUA Ala26-Pro653 (Ala26Thr) Accession # P35475.2
Formulation	Lyophilized from a 0.2 μm filtered solution in PBS and NaCl with Trehalose. See Certificate of Analysis for details. *Small pack size (-SP) is supplied either lyophilized or as a 0.2 μm filtered solution in PBS.

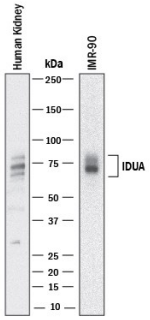
APPLICATIONS

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	2 µg/mL	See Below

DATA

Western Blot



Detection of Human α-L-Iduronidase/IDUA by Western Blot. Western blot shows lysates of human kidney tissue and IMR-90 human lung fibroblast cell line. PVDF membrane was probed with 2 µg/mL of Mouse Anti-Human α-L-Iduronidase/IDUA Monoclonal Antibody (Catalog # MAB4119) followed by HRP-conjugated Anti-Mouse IgG Secondary Antibody (Catalog # HAF018). A specific band was detected for α-L-Iduronidase/IDUA at approximately 74 kDa (as indicated). This experiment was conducted under reducing conditions and using Immunoblot Buffer Group 1.

PREPARATION AND STORAGE

Reconstitution	Reconstitute at 0.5 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.** • 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). The sequence of human IDUA shares 79% amino acid identity with mouse IDUA.

References:

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

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