

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
Conjugate	Alexa Fluor 594 Excitation Wavelength: 590 nm Emission Wavelength: 617 nm
Formulation	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.

Western Blot Optimal dilution of this antibody should be experimentally determined.

PREPARATION AND STORAGE

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

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.

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