SunLong Biotech Co.,LTD
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[Primary Antibody]

Rabbit Anti-IDUA antibody

Catalog Number: SL15542R

FOR RESEARCH USE ONLY

[DATASHEET]

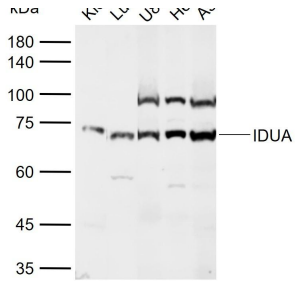
Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	IDUA
Immunogen:	KLH conjugated synthetic peptide derived from human IDUA: 101-200/653
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Applications:	WB=1:500-2000 ELISA=1:5000-10000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	Human, Mouse
Predicted MW.:	70kDa
Subcellular Location:	cytoplasmic
Background:	This gene encodes an enzyme that hydrolyzes the terminal alpha-L-iduronic acid residues of two glycosaminoglycans, dermatan sulfate and heparan sulfate. This hydrolysis is required for the lysosomal degradation of these glycosaminoglycans. Mutations in this gene that result in enzymatic deficiency lead to the autosomal recessive disease mucopolysaccharidosis type I (MPS I). [provided by RefSeq, Jul 2008]. **Subunit:** Monomer (Probable). **Subcellular Location:** Lysosome. **Tissue Specificity:** Ubiquitous. DISEASE: Mucopolysaccharidosis 1H (MPS1H) [MIM:607014]: A severe form of mucopolysaccharidosis type 1, a rare lysosomal storage disease characterized by progressive physical deterioration with urinary excretion of dermatan sulfate and heparan sulfate. Patients with MPS1H usually present, within the first year of life, a combination of hepatosplenomegaly, skeletal

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	deformities, corneal clouding and severe mental retardation. Obstructive airways disease, respiratory infection and cardiac complications usually result in death before 10 years of age. Note=The disease is caused by mutations affecting the gene represented in this entry. Mucopolysaccharidosis 1H/S (MPS1H/S) [MIM:607015]: A form of mucopolysaccharidosis type 1, a rare lysosomal storage disease characterized by progressive physical deterioration with urinary excretion of dermatan sulfate and heparan sulfate. MPS1H/S represents an intermediate phenotype of the MPS1 clinical spectrum. It is characterized by relatively little neurological involvement, but most of the somatic symptoms described for severe MPS1 develop in the early to mid-teens, causing considerable loss of mobility. Note=The disease is caused by mutations affecting the gene represented in this entry. Mucopolysaccharidosis 1S (MPS1S) [MIM:607016]: A mild form of mucopolysaccharidosis type 1, a rare lysosomal storage disease characterized by progressive physical deterioration with urinary excretion of dermatan sulfate and heparan sulfate. Patients with MPS1S may have little or no neurological involvement, normal stature and life span, but present development of joints stiffness, mild hepatosplenomegaly, aortic valve disease and corneal clouding. Note=The disease is caused by mutations affecting the gene represented in this entry. **Similarity:** Belongs to the glycosyl hydrolase 39 family. **Database links:** **Entrez Gene:** 3425 Human **Entrez Gene:** 15932 Mouse Omim: 252800 Human **SwissProt:** P35475 Human **SwissProt:** P48441 Mouse **Unigene:** 89560 Human

[VALIDATION IMAGES]



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Sample:
Lane 1: Mouse Kidney tissue lysates
Lane 2: Mouse Lung tissue lysates
Lane 3: Human U87MG cell lysates
Lane 4: Human HeLa cell lysates
Lane 5: Human A549 cell lysates
Primary: Anti-IDUA (SL15542R) at 1/1000 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 70 kDa
Observed band size: 70 kDa

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