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Rabbit Anti-IDUA antibody

SL15542R

Product Name:	IDUA
Chinese Name:	α-L-艾杜糖苷酶抗体
Alias:	IDUA_HUMAN; Alpha-L-iduronidase; IDA; Iduronidase alpha L; MPS1.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Horse,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	70kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human IDUA:101-200/653
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	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 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.

SWISS:
P35475

Gene ID:
3425

Database links:

Entrez Gene: 3425Human

	Entrez Gene: 15932Mouse Omim: 252800Human SwissProt: P35475Human SwissProt: P48441Mouse Unigene: 89560Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.