For Research Use Only

IDUA Polyclonal antibody

Catalog Number: 30006-1-AP



Basic Information

Catalog Number: 30006-1-AP

300 μg/ml

Isotype:

GenBank Accession Number:

NM 000203

GeneID (NCBI): Size:

UNIPROT ID: Source: Rabbit P35475 Full Name:

> iduronidase, alpha-L-Calculated MW:

Immunogen Catalog Number: AG30658 73 kDa

Observed MW:

73 kDa

Purification Method: Antigen affinity purification Recommended Dilutions: WB 1:500-1:3000 IHC 1:50-1:500

Applications

Tested Applications:

IHC, WB, ELISA

Species Specificity:

Human, Mouse, rat

Note-IHC: suggested antigen retrieval with TE buffer pH 9.0; (*) Alternatively, antigen retrieval may be performed with citrate

buffer pH 6.0

Positive Controls:

WB: A549 cells, HEK-293 cells, LNCaP cells, mouse

brain tissue, rat brain tissue

IHC: mouse kidney tissue,

Background Information

Iduronidase (L-iduronidase, alpha-L-iduronidase, laronidase) is an enzyme with the systematic name glycosaminoglycan alpha-L-iduronohydrolase. This enzyme catalyzes the hydrolysis of unsulfated alpha-Liduronosidic linkages in dermatan sulfate. It is a glycoprotein enzyme found in the lysosomes of cells. It is involved in the degeneration of glycosaminoglycans such as dermatan sulfate and heparan sulfate. The enzyme acts by hydrolyzing the terminal alpha-L-iduronic acid residues of these molecules, degrading them (PMID: 4993544,30407). A deficiency in the IDUA protein is associated with mucopolysaccharidoses (MPS). MPS, a type of lysosomal storage disease, is typed I through VII. In this syndrome, glycosaminoglycans accumulate in the lysosomes and cause substantial disease in many different tissues of the body. IDUA mutations result in the MPS 1 phenotype, which is $inherited \ in \ an \ autosomal \ recessive \ fashion. The \ defective \ alpha-L-iduroni dase \ results \ in \ an \ accumulation \ of \ alpha-L-iduroni dase \ results \ in \ an \ accumulation \ of \ alpha-L-iduroni dase \ results \ in \ an \ accumulation \ of \ alpha-L-iduroni dase \ results \ in \ an \ accumulation \ of \ alpha-L-iduroni dase \ results \ in \ an \ accumulation \ of \ alpha-L-iduroni dase \ results \ in \ an \ accumulation \ of \ alpha-L-iduroni dase \ results \ in \ an \ accumulation \ of \ alpha-L-iduroni dase \ results \ in \ an \ accumulation \ of \ alpha-L-iduroni dase \ results \ in \ an \ accumulation \ of \ alpha-L-iduroni dase \ results \ in \ an \ accumulation \ of \ a$ heparan and dermatan sulfate within phagocytes, endothelium, smooth muscle cells, neurons, and fibroblasts. Prenatal diagnosis of this enzyme deficiency is possible (PMID:8242073).

Storage

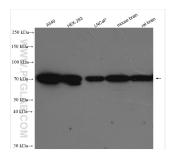
Store at -20°C. Stable for one year after shipment.

Storage Buffer:

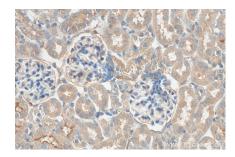
PBS with 0.02% sodium azide and 50% glycerol pH 7.3.

Aliquoting is unnecessary for -20°C storage

Selected Validation Data



Various lysates were subjected to SDS PAGE followed by western blot with 30006-1-AP (IDUA antibody) at dilution of 1:1500 incubated at room temperature for 1.5 hours.



Immunohistochemical analysis of paraffinembedded mouse kidney tissue slide using 30006-1-AP (IDUA antibody) at dilution of 1:200 (under 40x lens). Heat mediated antigen retrieval with Tris-EDTA buffer (pH 9.0).