己醛醣酸盐水解酵素抗体

产品货号： mlR18121

英文名称： IDUA

中文名称： 己醛醣酸盐水解酵素抗体

别　　名： Alpha L iduronidase; IDA; Iduronidase alpha L; MPS1.

研究领域： 肿瘤　细胞生物　免疫学　信号转导　细胞类型标志物

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 70kDa

细胞定位： 细胞浆

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human IDUA:561-653/653

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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

产品介绍： 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]

Function:

IDUA is 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. Defects in IDUA are the cause of mucopolysaccharidosis type 1H (MPS1H) also known as Hurler syndrome, mucopolysaccharidosis type 1H/S (MPS1H/S) also known as Hurler-Scheie syndrome and mucopolysaccharidosis type 1S (MPS1S) also known as Scheie syndrome. MPS1S is a mild form whilst MPS1H is a severe form of this rare lysosomal storage disease characterized by progressive physical deterioration with urinary excretion of dermatan sulfate and heparan sulfate.

Subunit:

Monomer.

Subcellular Location:

Lysosome

Tissue Specificity:

Ubiquitous.

Similarity:

Belongs to the glycosyl hydrolase 39 family.

SWISS:

P35475

Gene ID:

3425

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.