艾杜糖-2-硫酸酯酶抗体

产品货号： mlR15543

英文名称： Iduronate 2 sulfatase

中文名称： 艾杜糖-2-硫酸酯酶抗体

别　　名： Alpha L iduronate sulfate sulfatase; Alpha-L-iduronate sulfate sulfatase; AW214631; Ids; IDS_HUMAN; Iduronate 2 sulfatase 14 kDa chain; Iduronate 2 sulfatase 42 kDa chain; Iduronate 2 sulfatase; Iduronate 2-sulfatase 14 kDa chain; Iduronate sulfatase; Idursulfase; MPS2; RP23-29M4.1; SIDS.

研究领域： 细胞生物　免疫学　细胞类型标志物

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Dog, Cow, Sheep,

产品应用： WB=1:500-2000 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： 47kDa

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 Iduronate 2 sulfatase:101-200/550

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 ℃ 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℃. 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 ℃.

PubMed： PubMed

产品介绍： The protein encoded by this gene belongs to the sulfatase family, is localized to the lysosome, and is involved in lysosomal degradation of heparan sulfate and dermatan sulfate. Mutations in this gene are associated with the X-linked lysosomal storage disease, mucopolysaccharidosis type II, also known as Hunter syndrome. Alternatively spliced transcript variants have been described for this gene. [provided by RefSeq, Aug 2013]

Function:

Required for the lysosomal degradation of heparan sulfate and dermatan sulfate.

Subunit:

Liver iduronate 2-sulfatase is composed of two major forms (A and B) which contain both a 42 kDa and a 14 kDa polypeptides.

Subcellular Location:

Lysosome.

Tissue Specificity:

Liver, kidney, lung, and placenta.

Post-translational modifications:

The conversion to 3-oxoalanine (also known as C-formylglycine, FGly), of a serine or cysteine residue in prokaryotes and of a cysteine residue in eukaryotes, is critical for catalytic activity (By similarity).

DISEASE:

Mucopolysaccharidosis 2 (MPS2) [MIM:309900]: An X-linked lysosomal storage disease characterized by intracellular accumulation of heparan sulfate and dermatan sulfate and their excretion in urine. Most children with MPS2 have a severe form with early somatic abnormalities including skeletal deformities, hepatosplenomegaly, and progressive cardiopulmonary deterioration. A prominent feature is neurological damage that presents as developmental delay and hyperactivity but progresses to mental retardation and dementia. They die before 15 years of age, usually as a result of obstructive airway disease or cardiac failure. In contrast, those with a mild form of MPS2 may survive into adulthood, with attenuated somatic complications and often without mental retardation. Note=The disease is caused by mutations affecting the gene represented in this entry.

Similarity:

Belongs to the sulfatase family.

SWISS:

P22304

Gene ID:

3423

Important Note:

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

产品图片