

Rabbit Anti-Iduronate 2 sulfatase antibody

SL15543R

Product Name:	Iduronate 2 sulfatase
Chinese Name:	艾杜糖-2-硫酸酯酶抗体
Alias:	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.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human, Mouse, Rat, Dog, Cow, Sheep,
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:	47kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	lmg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Iduronate 2 sulfatase:101-200/550
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:	<u>PubMed</u>
Product Detail:	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

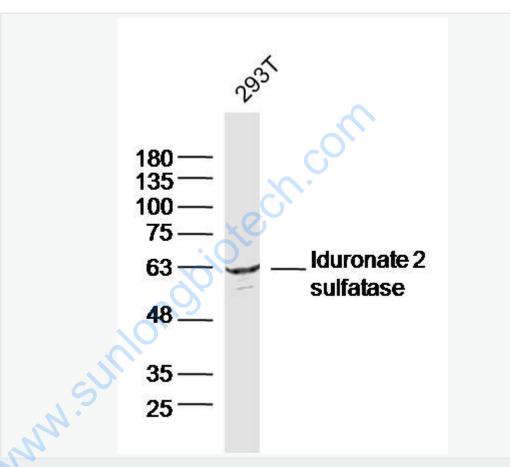
Database links:

Entrez Gene: 3423 Human Omim: 309900 Human SwissProt: P22304 Human Unigene: 460960 Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. 180 135 100 **75** — Iduronate 2 sulfatase Picture: 25 Sample: Hela Cell(Human)Lysate at 30 ug Primary: Anti- Iduronate 2 sulfatase (SL15543R)at 1/300 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 47kD

Observed band size: 62kD



Sample:

293T Cell (Human) Lysate at 30 ug

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