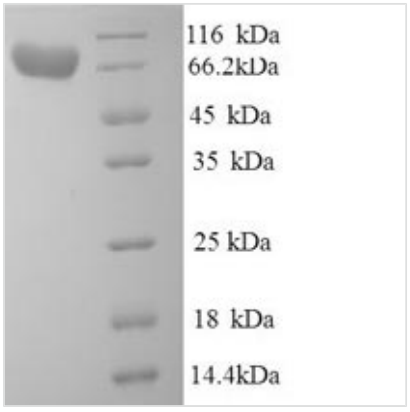
Recombinant Human Alpha-L-iduronidase (IDUA)

Product Code	CSB-YP011000HU
Abbreviation	Recombinant Human IDUA protein
Storage	The shelf life is related to many factors, storage state, buffer ingredients, storage temperature and the stability of the protein itself. Generally, the shelf life of liquid form is 6 months at -20°C/-80°C. The shelf life of lyophilized form is 12 months at -20°C/-80°C.
Uniprot No.	P35475
Product Type	Recombinant Protein
Immunogen Species	Homo sapiens (Human)
Purity	Greater than 90% as determined by SDS-PAGE.
Sequence	APHLVHVDAARALWPLRRFWRSTGFCPPLPHSQADQYVLSWDQQLNLAYVG AVPHRGIKQVRTHWLLELVTTRGSTGRGLSYNFTHLDGYLDLLRENQLLPGFE LMGSASGHFTDFEDKQQVFEWKDLVSSLARRYIGRYGLAHVSKWNFETWNE PDHHDFDNVSMTMQGFLNYYDACSEGLRAASPALRLGGPGDSFHTPPRSPL SWGLLRHCHDGTNFFTGEAGVRLDYISLHRKGARSSISILEQEKVVAQQIRQLF PKFADTPIYNDEADPLVGWSLPQPWRADVTYAAMVVKVIAQHQNLLLANTTSA FPYALLSNDNAFLSYHPHPFAQRTLTARFQVNNTRPPHVQLLRKPVLTAMGLL ALLDEEQLWAEVSQAGTVLDSNHTVGVLASAHRPQGPADAWRAAVLIYASDD TRAHPNRSVAVTLRLRGVPPGPGLVYVTRYLDNGLCSPDGEWRRLGRPVFPT AEQFRRMRAAEDPVAAAPRPLPAGGRLTLRPALRLPSLLLVHVCARPEKPPG QVTRLRALPLTQGQLVLVWSDEHVGSKCLWTYEIQFSQDGKAYTPVSRKPST FNLFVFSPDTGAVSGSYRVRALDYWARPGPFSDPVPYLEVPVPRGPPSPGNP
Research Area	Metabolism
Source	Yeast
Target Names	IDUA
Protein Names	Recommended name: Alpha-L-iduronidase EC= 3.2.1.76
Expression Region	28-653aa
Notes	Repeated freezing and thawing is not recommended. Store working aliquots at 4°C for up to one week.
Tag Info	N-terminal 6xHis-tagged
Mol. Weight	71.9kDa
Protein Length	Full Length of Mature Protein
Image	



(Tris-Glycine gel) Discontinuous SDS-PAGE (reduced) with 5% enrichment gel and 15% separation gel.

Description

The IDUA (alpha-L-iduronidase) protein is a lysosomal enzyme that exists as a dimer and is essential for the degradation of glycosaminoglycans (GAGs) such as dermatan sulfate and heparan sulfate [1]. Mutations affecting the enzyme activity of IDUA can lead to mucopolysaccharidosis type I (MPS I), a lysosomal storage disorder characterized by the accumulation of GAGs [2]. The IDUA enzyme activity is typically measured using a fluorometric enzymatic assay, which quantifies the conversion of 4-methylumbelliferyl-α-L-iduronide to 4-methylumbelliferone [3]. The specific activity of the IDUA enzyme is crucial, as it determines the effectiveness of therapeutic interventions. For instance, the high brain uptake of a fusion protein containing IDUA can lead to a significant normalization of brain IDUA enzyme activity following administration [4].

Furthermore, the IDUA protein is subject to genetic variations, and mutations affecting its activity have been identified in patients with MPS I [5]. These mutations can significantly reduce the enzyme activity of IDUA, leading to the manifestation of MPS I symptoms [5]. Additionally, the IDUA protein can be engineered into fusion proteins, such as the HIRMAb-IDUA fusion protein, to enhance its targeted delivery across the blood-brain barrier [6]. This targeted delivery is crucial for the treatment of neurological manifestations of MPS I, as it allows for the restoration of IDUA enzyme activity in the brain [7].

In summary, the IDUA protein is a critical lysosomal enzyme involved in the degradation of GAGs, and its enzyme activity is essential for maintaining cellular homeostasis. Mutations affecting the enzyme activity of IDUA can lead to the development of MPS I, highlighting the significance of understanding the protein's function and its potential therapeutic applications.

References:

[1] R. Boado, E. Hui, J. Lu, Q. Zhou, & W. Pardridge, "Reversal of lysosomal storage in brain of adult mps-i mice with intravenous trojan horse-iduronidase fusion protein", Molecular Pharmaceutics, vol. 8, no. 4, p. 1342-1350, 2011. https://doi.org/10.1021/mp200136x

[2] G. Lee-Chen, S. Lin, Y. Tang, & Y. Chin, "Mucopolysaccharidosis type i: characterization of novel mutations affecting α? l?iduronidase activity", Clinical Genetics, vol. 56, no. 1, p. 66-70, 1999. https://doi.org/10.1034/j.1399-0004.1999.560109.x

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[4] R. Boado, E. Hui, J. Lu, & W. Pardridge, "Agt-181: expression in cho cells and pharmacokinetics, safety, and plasma iduronidase enzyme activity in rhesus monkeys", Journal of Biotechnology, vol. 144, no. 2, p. 135-141, 2009. https://doi.org/10.1016/j.jbiotec.2009.08.019
[5] D. Liu, Z. Jiang, L. Deng, H. Li, & H. Jiang, "Identification of an α?l?iduronidase (idua) m1t mutation in a chinese family with autosomal recessive mucopolysaccharidosis i", Annals of the New York Academy of Sciences, vol. 1526, no. 1, p. 114-125, 2023. https://doi.org/10.1111/nyas.15016
[6] R. Boado, Y. Zhang, Y. Zhang, C. Xia, Y. Wang, & W. Pardridge, "Genetic engineering of a lysosomal enzyme fusion protein for targeted delivery across the human blood?brain barrier", Biotechnology and Bioengineering, vol. 99, no. 2, p. 475-484, 2007. https://doi.org/10.1002/bit.21602
[7] R. Boado and W. Pardridge, "Brain and organ uptake in the rhesus monkey in vivo of recombinant iduronidase compared to an insulin receptor antibody–iduronidase fusion protein", Molecular Pharmaceutics, vol. 14, no. 4, p. 1271-1277, 2017. https://doi.org/10.1021/acs.molpharmaceut.6b01166

Reconstitution	We recommend that this vial be briefly centrifuged prior to opening to bring the contents to the bottom. Please reconstitute protein in deionized sterile water to a concentration of 0.1-1.0 mg/mL.We recommend to add 5-50% of glycerol (final concentration) and aliquot for long-term storage at -20°C/-80°C. Our default final concentration of glycerol is 50%. Customers could use it as reference.
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