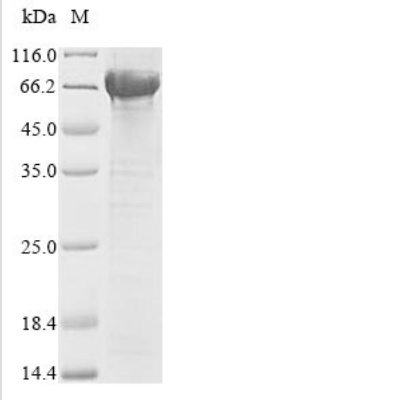
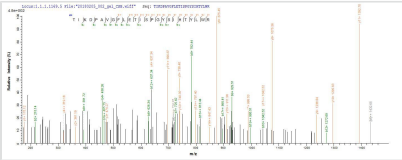
Recombinant Human Lysosomal acid glucosylceramidase (GBA1)

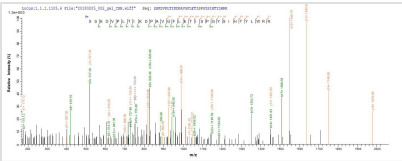
Product Code	CSB-EP009289HU
Abbreviation	Recombinant Human GBA protein
Storage	The shelf life is related to many factors, storage state, buffer ingredients, storage temperature and the stability of the protein itself. Generally, the shelf life of liquid form is 6 months at -20°C/-80°C. The shelf life of lyophilized form is 12 months at -20°C/-80°C.
Uniprot No.	P04062
Storage Buffer	Tris-based buffer,50% glycerol
Product Type	Recombinant Proteins
Immunogen Species	Homo sapiens (Human)
Purity	Greater than 85% as determined by SDS-PAGE.
Sequence	ARPCIPKSFGYSSVVCVCNATYCDSFDPPTFPALGTFSRYESTRSGRRMELS MGPIQANHTGTGLLLTLQPEQKFQKVKGFGGAMTDAAALNILALSPPAQNLLL KSYFSEEGIGYNIIRVPMASCDFSIRTYTYADTPDDFQLHNFSLPEEDTKLKIPLI HRALQLAQRPVSLLASPWTSPTWLKTNGAVNGKGSLKGQPGDIYHQTWARY FVKFLDAYAEHKLQFWAVTAENEPSAGLLSGYPFQCLGFTPEHQRDFIARDLG PTLANSTHHNVRLLMLDDQRLLLPHWAKVVLTDPEAAKYVHGIAVHWYLDFLA PAKATLGETHRLFPNTMLFASEACVGSKFWEQSVRLGSWDRGMQYSHSIITN LLYHVVGWTDWNLALNPEGGPNWVRNFVDSPIIVDITKDTFYKQPMFYHLGHF SKFIPEGSQRVGLVASQKNDLDAVALMHPDGSAVVVVLNRSSKDVPLTIKDPA VGFLETISPGYSIHTYLWRRQ
Research Area	Neuroscience
Source	E.coli
Target Names	GBA1
Protein Names	Recommended name: Glucosylceramidase EC= 3.2.1.45Alternative name(s): Acid beta-glucosidase Alglucerase Beta-glucocerebrosidase D-glucosyl-N-acylsphingosine glucohydrolase Imiglucerase
Expression Region	40-536aa
Notes	Repeated freezing and thawing is not recommended. Store working aliquots at 4°C for up to one week.
Tag Info	N-terminal 10xHis-SUMO-tagged and C-terminal Myc-tagged
Mol. Weight	75.6 kDa
Protein Length	Full Length of Mature Protein
Image	



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



Based on the SEQUEST from database of E.coli host and target protein, the LC-MS/MS Analysis result of CSB-EP009289HU could indicate that this peptide derived from E.coli-expressed Homo sapiens (Human) GBA.



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Description

To synthesize recombinant human lysosomal acid glucosylceramidase (GBA1), the target gene encoding the 40-536aa of human GBA1 is first isolated, with an N-terminal 10xHis-SUMO-tag and C-terminal Myc-tag. This gene is cloned into an expression vector, which is introduced into E. coli cells via transformation. The E. coli cells express the recombinant GBA1 protein, which is subsequently harvested from the cell lysate. The protein is purified using affinity chromatography. Finally, the protein's purity is determined by SDS-PAGE, reaching up to 85%.

The human GBA1 gene encodes the lysosomal enzyme beta-glucocerebrosidase (GCase), which plays a crucial role in glycosphingolipid substrate metabolism [1]. Mutations in GBA1 lead to GCase deficiency, resulting in the accumulation of glucosylceramide and subsequent lysosomal dysfunction [2]. These mutations are associated with Gaucher disease, a lysosomal storage disorder characterized by GlcCer accumulation [2]. GBA1 mutations are also linked to an increased risk for synucleinopathies such as Parkinson's disease and dementia with Lewy bodies [3][4]. GBA1 deficiency can accelerate the progression of pathology in models of synucleinopathies [5]. Furthermore, GBA1 mutations negatively affect physiological α-synuclein tetramers, leading to the destabilization of multimers and accumulation of monomers [6].

References:
[1] N. Polinski, T. Martinez, A. Gorodinsky, R. Gareus, M. Sasner, M. Herberthet al., Decreased glucocerebrosidase activity and substrate accumulation of glycosphingolipids in a novel gba1 d409v knock-in mouse model, Plos One, vol. 16, no. 6, p. e0252325, 2021. https://doi.org/10.1371/journal.pone.0252325

[2] E. Schejter, S. Bialik, A. Shkedy, V. Levin-Salomon, S. Levin-Zaidman, & A. Kimchi, Death by over-eating: the gaucher disease associated gene gba1, identified in a screen for mediators of autophagic cell death, is necessary for developmental cell death in drosophila midgut, Cell Cycle, vol. 16, no. 21, p. 2003-2010, 2017. https://doi.org/10.1080/15384101.2017.1380134
[3] S. Sardi, P. Singh, S. Cheng, L. Shihabuddin, & M. Schlossmacher, Mutant <i>gba1</i> expression and synucleinopathy risk: first insights from cellular and mouse models, Neurodegenerative Diseases, vol. 10, no. 1-4, p. 195-202, 2012. https://doi.org/10.1159/000335038
[4] S. Sardi, J. Clarke, C. Viel, M. Chan, T. Tamsett, C. Treleavenet al., Augmenting cns glucocerebrosidase activity as a therapeutic strategy for parkinsonism and other gaucher-related synucleinopathies, Proceedings of the National Academy of Sciences, vol. 110, no. 9, p. 3537-3542, 2013. https://doi.org/10.1073/pnas.1220464110
[5] D. Kim, H. Hwang, S. Choi, S. Kwon, S. Lee, J. Parket al., D409h gba1 mutation accelerates the progression of pathology in a53t α-synuclein transgenic mouse model, Acta Neuropathologica Communications, vol. 6, no. 1, 2018. https://doi.org/10.1186/s40478-018-0538-9
[6] S. Kim, S. Yun, S. Lee, G. Umanah, V. Bandaru, X. Yinet al., Gba1 deficiency negatively affects physiological α-synuclein tetramers and related multimers, Proceedings of the National Academy of Sciences, vol. 115, no. 4, p. 798-803, 2018. https://doi.org/10.1073/pnas.1700465115

Shelf Life

The shelf life is related to many factors, storage state, buffer ingredients, storage temperature and the stability of the protein itself.
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