abcam

Product datasheet

Recombinant Human HEXB protein ab198424

1 Image

Description

Product name	Recombinant Human HEXB protein
Purity	> 82 % SDS-PAGE. Affinity purified.
Expression system	Baculovirus infected Sf9 cells
Accession	P07686
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MELCGLGLPRPPMLLALLLATLLAAMLALLTQVALVVQVA EAARAPSVSA KPGPALWP LPLSVKMTPNLLHLAPENFYISHSPNSTAGPSCTLLEEAF RRYHGYIFGFYKWHHEPAE FQAKTQVQQLLVSITLQSECDAFPNISSD ESYTLLVKEPVAVLKANRVWGALRGLETFS QLVYQDSYGTFTINESTI IDSPRFSHRGILIDTSRHYLPVKIILKTLDAMAFNKFNVLHWH IV DDQ SFPYQSITFPELSNKGSYSLSHVYTPNDVRMVIEYARLRGIR VLPEFDTP GHTLSWG KGQKDLLTPCYSRQNKLDSFGPINPTLNTTYSFLTTFFKEI SEVFPDQFIHLGGDEVEFK CWESNPKIQDFMRQKGFGTDFKKLESFYI QKVLDIIATINKGSIVWQEVFDDKAKLAPGTI VEVWKDSAYPEELSRV TASGFPVILSAPWYLDLISYGQDWRKYYKVEPLDFGGTQK QK QLFIGG EACLWGEYVDATNLTPRLWPRASAVGERLWSSKDVRDM DDAYDRLTRHRC R MVERGIAAQPLYAGYCNHENMDYKDDDDKHHHHHH
Predicted molecular weight	65 kDa including tags
Tags	DDDDK tag C-Terminus

Specifications

Our **Abpromise guarantee** covers the use of **ab198424** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	SDS-PAGE
Form	Liquid

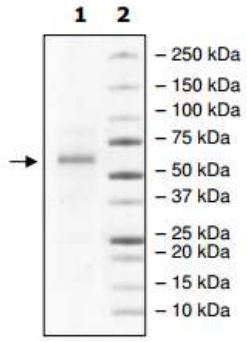
Preparation and Storage

Stability and Storage	Shipped on Dry Ice. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle. pH: 8.00 Preservative: 1.02% Imidazole Constituents: 0.63% Tris HCl, 0.64% Sodium chloride, 0.02% Potassium chloride, 0.04% Tween, 20% Glycerol (glycerin, glycerine)

General Info

Function	Responsible for the degradation of GM2 gangliosides, and a variety of other molecules containing terminal N-acetyl hexosamines, in the brain and other tissues.
Involvement in disease	Defects in HEXB are the cause of GM2-gangliosidosis type 2 (GM2G2) [MIM:268800]; also known as Sandhoff disease. GM2-gangliosidosis is an autosomal recessive lysosomal storage disease marked by the accumulation of GM2 gangliosides in the neuronal cells. GM2G2 is clinically indistinguishable from GM2-gangliosidosis type 1, presenting startle reactions, early blindness, progressive motor and mental deterioration, macrocephaly and cherry-red spots on the macula.
Sequence similarities	Belongs to the glycosyl hydrolase 20 family.
Post-translational modifications	N-linked glycans at Asn-142 and Asn-190 consist of Man(3)-GlcNAc(2) and Man(5 to 7)-GlcNAc(2), respectively. The beta-A and beta-B chains are produced by proteolytic processing of the precursor beta chain.
Cellular localization	Lysosome.

Images



SDS-PAGE - Recombinant Human HEXB protein (ab198424)

4-20% SDS-PAGE Coomassie staining.

Lane 1: 2.8 µg ab198424

Lane 2: Protein Marker

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