abcam

Product datasheet

Recombinant Human GNS protein ab158555

1 Image

Overview

Product name	Recombinant Human GNS protein
Protein length	Full length protein

Description

Nature	Recombinant
Source	Wheat germ
Amino Acid Sequence	
Species	Human
Sequence	MRLLPLAPGRLRRGSPRHLPSCSPALLLLVLGGCLGV FGVAAGTRRPNVV LLLTDDQDEVLGGMTPLKKTKALIGEMGMTFSSAYVP SALCCPSRASILT GKYPHNHHVVNNTLEGNCSSKSWQKIQEPNTFPAILRS MCGYQTFFAGKY LNEYGAPDAGGLEHVPLGWSYWYALEKNSKYYNYTLSI NGKARKHGENYS VDYLTDVLANVSLDFLDYKSNFEPFFMMIATPAPHSP WTAAPQYQKAFQN VFAPRNKNFNIHGTNKHWLIRQAKTPMTNSSIQFLDNA FRKRWQTLLSVD DLVEKLVKRLEFTGELNNTYIFYTSDNGYHTGQFSLPID KRQLYEFDIKV PLLVRGPGIKPNQTSKMLVANIDLGPTILDIAGYDLNKTQ MDGMSLLPIL RGASNLTWRSDVLVEYQGEGRNVTDPTCPSLSPGVS QCFPDCVCEDAYNN TYACVRTMSALWNLQYCEFDDQEVFVEVYNLTADPD QITNIAKTIDPELL GKMNYRLMMLQSCSGPTCRTPGVFDPGYRFDPRLMF SNRGSVRTRRFSKH LL
Amino acids	1 to 552
Tags	proprietary tag N-Terminus

Specifications

Our Abpromise guarantee covers the use of **ab158555** in the following tested applications.

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

Applications	Western blot ELISA
Form	Liquid
Additional notes	Protein concentration is above or equal to 0.05 mg/ml.

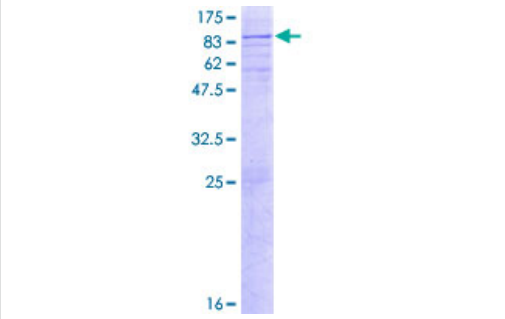
Preparation and Storage

Stability and Storage	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.31% Glutathione, 0.79% Tris HCl

General Info

Involvement in disease	Defects in GNS are the cause of mucopolysaccharidosis type 3D (MPS3D) [MIM:252940]; also known as Sanfilippo D syndrome. MPS3D is a form of mucopolysaccharidosis type 3, an autosomal recessive lysosomal storage disease due to impaired degradation of heparan sulfate. MPS3 is characterized by severe central nervous system degeneration, but only mild somatic disease. Onset of clinical features usually occurs between 2 and 6 years; severe neurologic degeneration occurs in most patients between 6 and 10 years of age, and death occurs typically during the second or third decade of life.
Sequence similarities	Belongs to the sulfatase family.
Post-translational modifications	The form A (78 kDa) is processed by internal peptidase cleavage to a 32 kDa N-terminal species (form B) and a 48 kDa C-terminal species. 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.
Cellular localization	Lysosome.

Images



SDS-PAGE - Recombinant Human GNS protein (ab158555)

ab158555 on a 12.5% SDS-PAGE stained with Coomassie Blue.

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