

Product datasheet

Recombinant Human PUS1 protein ab164258

1 Image

Overview

Product name	Recombinant Human PUS1 protein
Protein length	Full length protein

Description

Nature	Recombinant
Source	Wheat germ
Amino Acid Sequence	
Species	Human
Sequence	<p>MAGNAEPPPAGAACPQDRRSCSGRAGGDRVWEDG EHPAKKLKSGGDEERR EKPPKRKMLLMAYSGKGYHGMQRNVGSSQFKTIEDD LVSALVRSGCIPE NHGEDMRKMSFQRCARTDKGVSAAGQVVSLKVWLID DILEKINSHLPSHI RILGLKRVTGGFNSKNRCDARTYCYLLPTFAFAHKDRD VQDETYRLSAET LQQVNRLACYKGTNHNFTSQKGPQDPSACRYILE MYCEEPFVREGLE FAVIRVKGQSFMMHQIRKMVGLVVAIVKGYAPESVLER SWGTEKVDVPKA PGLGLVLERVHFEKYNQRFNGDGLHEPLDWAQEEGK VAAFKEEHYPTII GTERDERSMAQWLSTLPIHNFSATALTAGGTGAKVPS PLEGSEGDGDTD</p>
Amino acids	1 to 399
Tags	proprietary tag N-Terminus

Specifications

Our [Abpromise guarantee](#) covers the use of **ab164258** 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
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	ELISA
Form	Liquid
Additional notes	Protein concentration is above or equal to 0.05 mg/ml.

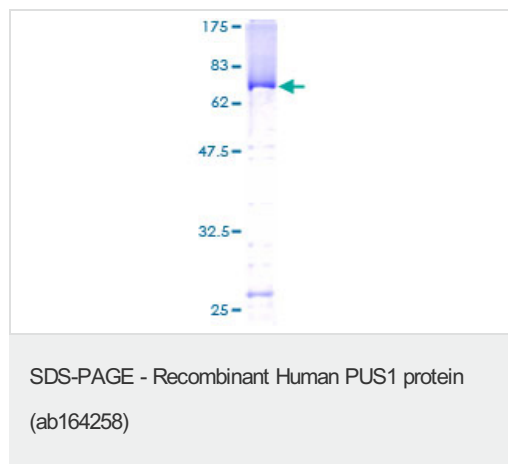
Preparation and Storage

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

Function	Converts specific uridines to PSI in a number of tRNA substrates. Acts on positions 27/28 in the anticodon stem and also positions 34 and 36 in the anticodon of an intron containing tRNA. Involved in regulation of nuclear receptor activity possibly through pseudouridylation of SRA1 RNA.
Tissue specificity	Widely expressed. High levels of expression found in brain and skeletal muscle.
Involvement in disease	Defects in PUS1 are a cause of myopathy with lactic acidosis and sideroblastic anemia type 1 (MLASA1) [MIM:600462]; also known as mitochondrial myopathy and sideroblastic anemia. MLASA is a rare autosomal recessive oxidative phosphorylation disorder specific to skeletal muscle and bone marrow.
Sequence similarities	Belongs to the tRNA pseudouridine synthase TruA family.
Cellular localization	Mitochondrion and Nucleus.

Images



ab164258 on a 12.5% SDS-PAGE stained with Coomassie Blue.

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