

Recombinant Human PUS1 protein ab164258

1 Image

Description	
Product name	Recombinant Human PUS1 protein
Expression system	Wheat germ
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MAGNAEPPPAGAACPQDRRSCSGRAGGDRVWEDGEHP AKKLKSGGDEERR EKPPKRKIVLLMAYSGKGYHGMQRNVGSSQFKTIEDDLVS ALVRSGCIPE NHGEDMRKMSFQRCARTDKGVSAAGQVVSLKVWLIDDIL EKINSHLP SHI RILGLKRVTTGGFNSKNRCDARTYCYLLPTFAFAHKDRDVQ DETYRLSAET LQQVNRLLACYKGTHNFHNFTSQKGPQDPSACRYILEMYC EPPFVREGLE FAVIRVKGQSFMMHQIRKMVGLVVAIVKGYAPESVLER SW GTEKVDVPKA PGLGLVLERVHFEKYNQRF GNDGLHEPLDWAQEEGKVA AFKEEHYPTII GTERDERSMAQWLSTLPIHNFSATALTAGGTGAKVPSPLE GSEGDGDTD
Amino acids	1 to 399
Tags	GST 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 ELISA
Form	Liquid

Additional notes

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

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.

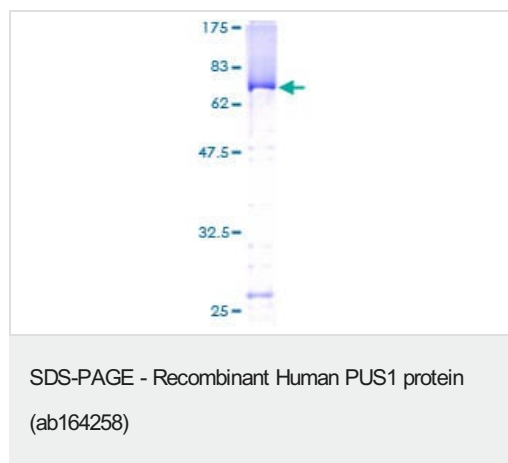
Sequence similarities

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.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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