abcam

Product datasheet

Recombinant Human POLG protein ab196066

1 Image

Description

Product name	Recombinant Human POLG protein
Purity	>= 81 % SDS-PAGE. Affinity purified.
Expression system	Baculovirus infected Sf9 cells
Accession	**P54098**
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MHHHHHHDYKDDDDKSRLLWRKVAGATVGPGPVPAPGR WVSSSVPASDPS DGQRR RQQQQQQQQQQQQQPQQPQVLSSEGGQLRHNPLDIQML SRGLH EQIFGQGGEMP GEAAVRRSVEHLQKHGLWGQPAVPLPDVELRLPPLYG DNLDQHFRLLAQKQSLPYL EAANLLLQAQLPPKPPAWAWAEGWTRYGP EGEAVPVAIPEERALVFDVEVCLAEGT CPTLAVAISPSAWYSWCSQRL VEERYSWTSQLSPADLIPLEVPTGASSPTQRDWQEQ LVVGHNVSFDRA HIREQYLIQGSRMRFLDTMSMHMAISGLSSFQRSLWIAAKQ GKHK VQP PTKQGQKSQRKARRGPAISSWDWLDISSVNSLAEVHRLY VGGPPLEKEPR ELFV KGTMKDIRENFQDLMQYCAQDVWATHEVFQQQLPLFLER CPHPV TLAGMLEMGVSY LPVNQNWERYLAEAQGTYEELQREMKKSLMDLANDA CQLLSGERYKEDPWLWDLE WDLQEFKQKKAKKVKKEPATASKLPIEGA GAPGDPMDQEDLGPCSEEEEFQQDVM ARACLQKLKGTTELLPKRPQHL PGHPGWYRKLCPRLDDPAWTPGPSLLSLQMRVTP KLMALTWDGFPLHY SERHGWGYLVPGRRDNLAKLPTGTTLESAGVVCPYRAIES

LYR KHCLE
QGKQQLMPQEAGLAEEFLLTDNSAIWQTVEELDYLEVEA
EAKMENLRAAV P
GQPLALTARGGPKDTQPSYHHGNGPYNDVDIPGCWFFKL
PHKDGNSC NVGSPFAKD
FLPKMEDGTLQAGPGGASGPRALEINKMISFWRNAHKRI
SSQMVVWLPRSALPRAVI
RHPDYDEEGLYGAILPQVVTAGTITRRAVE
PTWLTASNARPDRVGSELKAMVQAPPG
YTLVGADVDSQELWIAAVLGD
AHFAGMHGCTAFGWMTLQGRKSRGTDLHSKTATTV
GISREHAKIFNYG
RIYGAGQPFAERLLMQFNHRLTQQEAAEKAQQMYAATKG
LRWY RLSDE
GEWLVRELNLPVDRTEGGWISLQDLRKVQRETARKSQW
KKWEVVAERAWK
GGTESEMFNKLESIATSDIPRTPVLGCCISRALEPSAVQEE
FMTSRVN WVVQSSAVDY
LHLMLVAMKWLFEEFAIDGRFCISIHDEVRYLVREEDR
YRAALALQITNLLTRCMFAYK
LGLNDLPQSVAFFSAVDIDRCLRKEVT
MDCKTPSNPTGMERRYGIPQGEALDIYQIIEL
TKGSLEKRSQPGP

Predicted molecular weight	141 kDa including tags
Amino acids	2 to 1239
Tags	His tag N-Terminus , DDDDK tag N-Terminus
Additional sequence information	(NM_002693)

Specifications

Our **Abpromise guarantee** covers the use of **ab196066** 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
Additional notes	This product was previously labelled as DNA Polymerase gamma

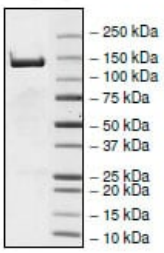
Preparation and Storage

Stability and Storage	Shipped on Dry Ice. Store at -80°C. Avoid freeze / thaw cycle. pH: 8.00 Constituents: 0.63% Tris HCl, 0.64% Sodium chloride, 0.02% Potassium chloride, 0.04% Tween, 20% Glycerol (glycerin, glycerine) also contains 80 µg/ml DDDDK peptide

General Info

Function	Involved in the replication of mitochondrial DNA.
Involvement in disease	Defects in POLG are the cause of progressive external ophthalmoplegia with mitochondrial DNA deletions autosomal dominant type 1 (PEOA1) [MIM:157640]. Progressive external ophthalmoplegia is characterized by progressive weakness of ocular muscles and levator muscle of the upper eyelid. In a minority of cases, it is associated with skeletal myopathy, which predominantly involves axial or proximal muscles and which causes abnormal fatigability and even permanent muscle weakness. Ragged-red fibers and atrophy are found on muscle biopsy. A large proportion of chronic ophthalmoplegias are associated with other symptoms, leading to a multisystemic pattern of this disease. Additional symptoms are variable, and may include cataracts, hearing loss, sensory axonal neuropathy, ataxia, depression, hypogonadism, and parkinsonism. Defects in POLG are a cause of progressive external ophthalmoplegia with mitochondrial DNA deletions autosomal recessive (PEOB) [MIM:258450]. PEOB is a severe form of progressive external ophthalmoplegia. It is clinically more heterogeneous than the autosomal dominant forms. Can be more severe. Defects in POLG are a cause of sensory ataxic neuropathy dysarthria and ophthalmoparesis (SANDO) [MIM:607459]. SANDO is a clinically heterogeneous systemic disorder with variable features resulting from mitochondrial dysfunction. It shares phenotypic characteristics with autosomal recessive progressive external ophthalmoplegia and mitochondrial neurogastrointestinal encephalopathy syndrome. The clinical triad of symptoms consists of sensory ataxic, neuropathy, dysarthria, and ophthalmoparesis. Defects in POLG are a cause of Alpers-Huttenlocher syndrome (AHS) [MIM:203700]; also called Alpers diffuse degeneration of cerebral gray matter with hepatic cirrhosis. AHS is an autosomal recessive hepatocerebral syndrome. The typical course of AHS includes severe developmental delay, intractable seizures, liver failure, and death in childhood. Refractory seizures, cortical blindness, progressive liver dysfunction, and acute liver failure after exposure to valproic acid are considered diagnostic features. The neuropathological hallmarks of AHS are neuronal loss, spongiform degeneration, and astrocytosis of the visual cortex. Liver biopsy results show steatosis, often progressing to cirrhosis. Defects in POLG are a cause of mitochondrial neurogastrointestinal encephalopathy syndrome (MNGIE) [MIM:603041]; also known as myoneurogastrointestinal encephalomyopathy. MNGIE is an autosomal recessive disease associated with multiple deletions of skeletal muscle mitochondrial DNA (MtDNA). It is clinically characterized by onset between the second and fifth decades of life, ptosis, progressive external ophthalmoplegia, gastrointestinal dysmotility (often pseudoobstruction), diffuse leukoencephalopathy, thin body habitus, peripheral neuropathy, and myopathy. Defects in POLG are a cause of Leigh syndrome (LS) [MIM:256000]. LS is a severe neurological disorder characterized by bilaterally symmetrical necrotic lesions in subcortical brain regions.
Sequence similarities	Belongs to the DNA polymerase type-A family.
Cellular localization	Mitochondrion.

Images



SDS-PAGE - Recombinant Human POLG protein (ab196066)

SDS-PAGE analysis of 1.3 µg ab196066 using 4-20 % SDS-PAGE gel and stained with Coomassie.

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