# abcam

# Product datasheet

# Recombinant Human PAF-1 protein ab159291

# 1 Image

**Description** 

Product name Recombinant Human PAF-1 protein

**Expression system** Wheat germ

Protein length Full length protein

Animal free No

**Nature** Recombinant

**Species** Human

**Sequence** MASRKENAKSANRVLRISQLDALELNKALEQLVWSQFTQ

CFHGFKPGLLA

RFEPEVKACLWVFLWRFTIYSKNATVGQSVLNIKYKNDFS

**PNLRYQPPSK** 

NQKIWYAVCTIGGRWLEERCYDLFRNHHLASFGKVKQCV

NFVIGLLKLGG

 ${\it LINFLIFLQRGKFATLTERLLGIHSVFCKPQNIReVGFEYMN}$ 

RELLWHGF

AEFLIFLLPLINVQKLKAKLSSWCIPLTGAPNSDNTLATSGK

**ECALCGEW** 

PTMPHTIGCEHIFCYFCAKSSFLFDVYFTCPKCGTEVHSLQ

PLKSGIEMS EVNAL

Amino acids 1 to 305

Tags GST tag N-Terminus

#### **Specifications**

Our Abpromise guarantee covers the use of ab159291 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 This product was previously labelled as PEX2.

#### **Preparation and Storage**

## **Stability and Storage**

Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.

00.8 :Hg

Constituents: 0.31% Glutathione, 0.79% Tris HCI

#### **General Info**

#### **Function**

Somewhat implicated in the biogenesis of peroxisomes.

#### Involvement in disease

Defects in PEX2 are the cause of peroxisome biogenesis disorder complementation group 5 (PBD-CG5) [MIM:170993]; also known as PBD-CGF. PBD refers to a group of peroxisomal disorders arising from a failure of protein import into the peroxisomal membrane or matrix. The PBD group is comprised of four disorders: Zellweger syndrome (ZWS), neonatal adrenoleukodystrophy (NALD), infantile Refsum disease (IRD), and classical rhizomelic chondrodysplasia punctata (RCDP). ZWS, NALD and IRD are distinct from RCDP and constitute a clinical continuum of overlapping phenotypes known as the Zellweger spectrum. The PBD group is genetically heterogeneous with at least 14 distinct genetic groups as concluded from complementation studies.

Defects in PEX2 are a cause of Zellweger syndrome (ZWS) [MIM:214100]. ZWS is a fatal peroxisome biogenesis disorder characterized by dysmorphic facial features, hepatomegaly, ocular abnormalities, renal cysts, hearing impairment, profound psychomotor retardation, severe hypotonia and neonatal seizures. Death occurs within the first year of life.

Defects in PEX2 are a cause of infantile Refsum disease (IRD) [MIM:266510]. IRD is a mild peroxisome biogenesis disorder (PBD). Clinical features include early onset, mental retardation, minor facial dysmorphism, retinopathy, sensorineural hearing deficit, hepatomegaly, osteoporosis, failure to thrive, and hypocholesterolemia. The biochemical abnormalities include accumulation of phytanic acid, very long chain fatty acids (VLCFA), di- and trihydroxycholestanoic acid and pipecolic acid.

#### Sequence similarities

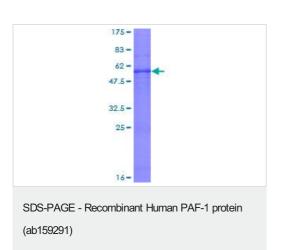
Belongs to the pex2/pex10/pex12 family.

Contains 1 RING-type zinc finger.

#### **Cellular localization**

Peroxisome membrane.

#### **Images**



ab159291 on a 12.5% SDS-PAGE stained with Coomassie Blue.

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