abcam

Product datasheet

Recombinant Human ENPP1/PC1 protein ab167943

1 References 1 Image

Description

Product name Recombinant Human ENPP1/PC1 protein

Purity > 95 % Densitometry.

ab167943 was purifed using Ni-NTA chromatography.

Endotoxin level < 1.000 Eu/μg
Expression system HEK 293 cells

Accession P22413

Protein length Protein fragment

Animal free No

Nature Recombinant

Species Human

Sequence

ASKPSCAKEV KSCKGRCFER TFGNCRCDAA CVELGNCCLD YQETCIEPEH IWTCNKFRCG EKRLTRSLCA CSDDCKDKGD CCINYSSVCQ GEKSWVEEPC ESINEPQCPA GFETPPTLLF SLDGFRAEYL HTWGGLLPVI SKLKKCGTYT

 ${\sf KNMRPVYPTK\ TFPNHYSIVT\ GLYPESHGII\ DNKMYDPKMN\ }$

ASFSLKSKEK FNPEWYKGEP IWVTAKYQGL

KSGTFFWPGS DVEINGIFPD IYKMYNGSVP FEERILAVLQ

WLQLPKDERP HFYTLYLEEP DSSGHSYGPV SSEVIKALQR VDGMVGMLMD GLKELNLHRC

LNLILISDHG MEQGSCKKYI YLNKYLGDVK NIKVIYGPAA

RLRPSDVPDK YYSFNYEGIA RNLSCREPNQ HFKPYLKHFL PKRLHFAKSD RIEPLTFYLD PQWQLALNPS ERKYCGSGFH GSDNVFSNMQ

ALFVGYGPGF KHGIEADTFE NIEVYNLMCD LLNLTPAPNN

GTHGSLNHLL KNPVYTPKHP KEVHPLVQCP

FTRNPRDNLG CSCNPSILPI EDFQTQFNLT VAEEKIIKHE

TLPYGRPRVL QKENTICLLS QHQFMSGYSQ DILMPLWTSY TVDRNDSFST EDFSNCLYQD

FRIPLSPVHK CSFYKNNTKV SYGFLSPPQL NKNSSGIYSE ALLTTNIVPM YQSFQVIWRY FHDTLLRKYA EERNGVNVVS GPVFDFDYDG RCDSLENLRQ KRRVIRNQEI LIPTHFFIVL

TSCKDTSQTP LHCENLDTLA FILPHRTDNS ESCVHGKHDS SWVEELLMLH RARITDVEHI

TGLSFYQQRK EPVSDILKLK THLPTFSQED GPKLHHHHHH

Predicted molecular weight 97 kDa including tags

Amino acids 98 to 925

Tags His tag C-Terminus

Specifications

Our Abpromise guarantee covers the use of ab167943 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

SDS-PAGE

Form Lyophilized

Additional notes This product was previously labelled as ENPP1

Preparation and Storage

Stability and Storage Shipped at 4°C. Store at -80°C.

Constituents: 99% Phosphate Buffer, 0.43% Sodium chloride

Reconstitution Add 200µl deionized water to prepare a working stock solution of 0.5 mg/mL and let the

lyophilized pellet dissolve completely. Aliquot reconstituted protein to avoid repeated

freezing/thawing cycles and store at -80°C for long term storage.

Product is not sterile! Please filter the product by an appropriate sterile filter before using it in the

cell culture.

General Info

Function Involved primarily in ATP hydrolysis at the plasma membrane. Plays a role in regulating

pyrophosphate levels, and functions in bone mineralization and soft tissue calcification. In vitro, has a broad specificity, hydrolyzing other nucleoside 5' triphosphates such as GTP, CTP, TTP and UTP to their corresponding monophosphates with release of pyrophosphate and diadenosine polyphosphates, and also 3',5'-cAMP to AMP. May also be involved in the regulation of the availability of nucleotide sugars in the endoplasmic reticulum and Golgi, and the regulation of

purinergic signaling. Appears to modulate insulin sensitivity.

Tissue specificity Expressed in plasma cells and also in a number of non-lymphoid tissues, including the distal

convoluted tubule of the kidney, chondrocytes and epididymis.

Involvement in disease Defects in ENPP1 are a cause of increased susceptibility for ossification of the posterior

longitudinal ligament of the spine (OPLL) [MIM:602475]. OPLL is a common form of human

myelopathy with a prevalence of as much as 4% in a variety of ethnic groups.

Defects in ENPP1 are the cause of arterial calcification of infancy, generalized, type 1 (GACI1) [MIM:208000]. A severe autosomal recessive disorder characterized by calcification of the internal elastic lamina of muscular arteries and stenosis due to myointimal proliferation. The disorder is often fatal within the first 6 months of life because of myocardial ischemia resulting in

refractory heart failure.

Defects in ENPP1 are associated with obesity, glucose intolerance, and type II diabetes non-insulin dependent (NIDDM) [MIM:125853].

Defects in ENPP1 are the cause of rickets hypophosphatemic autosomal recessive type 2 (ARHR2) [MIM:613312]. ARHR2 is a hereditary form of hypophosphatemic rickets, a disorder of proximal renal tubule function that causes phosphate loss, hypophosphatemia and skeletal deformities, including rickets and osteomalacia unresponsive to vitamin D. Symptoms are bone pain, fractures and growth abnormalities.

Sequence similarities

Belongs to the nucleotide pyrophosphatase/phosphodiesterase family.

Contains 2 SMB (somatomedin-B) domains.

Domain

The di-leucine motif is required for basolateral targeting in epithelial cells, and for targeting to

matrix vesicles derived from mineralizing cells.

Post-translational modifications

Autophosphorylated as part of the catalytic cycle of phosphodiesterase/pyrophosphatase activity.

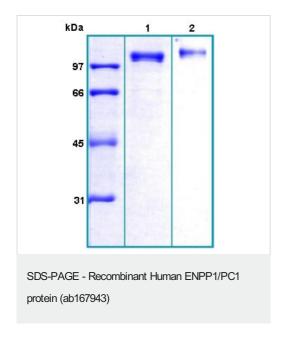
N-glycosylated.

It has been suggested that the active SMB domain may be permitted considerable disulfide bond heterogeneity or variability, thus two alternate disulfide patterns based on 3D structures are described with 1 disulfide bond conserved in both.

Cellular localization

Membrane. Basolateral cell membrane. Targeted to the basolateral membrane in polarized epithelial cells and in hepatocytes, and to matrix vesicles in osteoblasts. In bile duct cells and cancer cells, located to the apical cytoplasmic side.

Images



10% SDS-PAGE analysis of ab167943.

Lane 1: reduced and boiled sample, 2.5µg/lane.

Lane 2: non-reduced and non-boiled sample, 2.5µg/lane.

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