Product datasheet

Anti-ENPP1/PC1 antibody ab40003

2 Abreviews 7 References 1 Image

Overview

Product name Anti-ENPP1/PC1 antibody
Description Goat polyclonal to ENPP1/PC1
Host species Goat
Tested applications Suitable for: IHC-Fr, IHC-P
Species reactivity Reacts with: Human
Predicted to work with: Mouse, Rat
Immunogen Synthetic peptide corresponding to Human ENPP1/PC1 aa 915-925 (C terminal).
Sequence: KTHLPTFSQED
Positive control Human liver and pancreas tissue sections. Lysate prepared from transfected HEK293 cells transiently expressing ENPP1/PC1.
General notes This product was previously labelled as ENPP1

Properties

Form Liquid
Storage instructions Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
Storage buffer pH: 7.30
Preservative: 0.02% Sodium azide
 Constituents: Tris buffered saline, 0.5% BSA
Purity Immunogen affinity purified
Purification notes Purified from goat serum by ammonium sulphate precipitation followed by antigen affinity chromatography using the immunizing peptide.
Clonality Polyclonal
Isotype IgG

Applications

Run BLAST with
Run BLAST with
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.

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Our Abpromise guarantee covers the use of ab40003 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

<table>
<thead>
<tr>
<th>Application</th>
<th>Abreviews</th>
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<tbody>
<tr>
<td>IHC-Fr</td>
<td>1/250. PubMed: 23225236</td>
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<tr>
<td>IHC-P</td>
<td>Use a concentration of 2 - 4 µg/ml</td>
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Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-ENPP1/PC1 antibody (ab40003)

Ab40003 (2 μg/ml) staining human ENPP1/PC1 in human liver tissue by immunohistochemistry using paraffin embedded tissue. Microwaved antigen retrieval with citrate buffer pH 6, HRP-staining.

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