

DPP1 Rabbit pAb

Catalog # AP98540

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P53634
Other Accession	P53634
Host	Rabbit
Clonality	Polyclonal
Calculated MW	51854
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human cathepsin C light chain
Epitope Specificity	365-463/463
Isotype	IgG
Purity	affinity purified by Protein A

Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the peptidase C1 family.
SUBUNIT	Tetramer of heterotrimers consisting of exclusion domain, heavy- and light chains.
Post-translational modifications	N-glycosylated. While glycosylation at Asn-53, Asn-119 and Asn-276 is mediated by STT3A-containing complexes, glycosylation at Asn-29 is mediated STT3B-containing complexes. In approximately 50% of the complexes the exclusion domain is cleaved at position 58 or 61. The two parts of the exclusion domain are held together by a disulfide bond.
DISEASE	Papillon-Lefevre syndrome (PLS) [MIM:245000]: An autosomal recessive disorder characterized by palmoplantar keratosis and severe periodontitis affecting deciduous and permanent dentitions and resulting in premature tooth loss. The palmoplantar keratotic phenotype vary from mild psoriasiform scaly skin to overt hyperkeratosis. Keratosis also affects other sites such as elbows and knees. Note=The disease is caused by mutations affecting the gene represented in this entry. Haim-Munk syndrome (HMS) [MIM:245010]: An autosomal recessive disorder characterized by palmoplantar keratosis, onychogryphosis and periodontitis. Additional features are pes planus, arachnodactyly, and acroosteolysis. Note=The disease is caused by mutations affecting the gene represented in this entry. Periodontitis, aggressive, 1 (AP1) [MIM:170650]: A disease characterized by severe and protracted gingival infections, generalized or localized, leading to tooth loss. Amounts of microbial deposits are generally inconsistent with the severity of periodontal tissue destruction and the progression of attachment and bone loss may be self arresting. Note=The disease is caused by mutations affecting the gene represented in this entry.

Important Note This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Background Descriptions The protein encoded by this gene, a member of the peptidase C1 family, is a lysosomal cysteine proteinase that appears to be a central coordinator for activation of many serine proteinases in immune/inflammatory cells. It is composed of a dimer of disulfide-linked heavy and light chains, both produced from a single protein precursor, and a residual portion of the propeptide acts as an intramolecular chaperone for the folding and

stabilization of the mature enzyme. This enzyme requires chloride ions for activity and can degrade glucagon. Defects in the encoded protein have been shown to be a cause of Papillon-Lefevre syndrome, an autosomal recessive disorder characterized by palmoplantar keratosis and periodontitis. Multiple transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq].

Additional Information

Gene ID	1075
Other Names	CPPI; DPP-I; DPP1; DPPI; HMS; JP; JPD; PALS; PDON1; PLS; CATC_HUMAN; CTSC; Cathepsin C; Cathepsin J; Dipeptidyl peptidase I (DPP-I DPPI); Dipeptidyl transferase; 3.4.14.1; dipeptidyl peptidase 1
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	CTSC
Synonyms	CPPI
Function	Thiol protease (PubMed: 1586157). Has dipeptidylpeptidase activity (PubMed: 1586157). Active against a broad range of dipeptide substrates composed of both polar and hydrophobic amino acids (PubMed: 1586157). Proline cannot occupy the P1 position and arginine cannot occupy the P2 position of the substrate (PubMed: 1586157). Can act as both an exopeptidase and endopeptidase (PubMed: 1586157). Activates serine proteases such as elastase, cathepsin G and granzymes A and B (PubMed: 8428921).
Cellular Location	Lysosome.
Tissue Location	Ubiquitous. Highly expressed in lung, kidney and placenta. Detected at intermediate levels in colon, small intestine, spleen and pancreas.

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