



Dipeptidyl-peptidase 1 (heavy chain, Cleaved-Leu231) rabbit pAb

Cat No.:ES19987

For research use only

Overview

Product Name	Dipeptidyl-peptidase 1 (heavy chain, Cleaved-Leu231) rabbit pAb
Host species	Rabbit
Applications	WB; ELISA
Species Cross-Reactivity	Human;Rat;Mouse;
Recommended dilutions	WB 1:1000-2000 ELISA 1:5000-20000
Immunogen	Synthesized peptide derived from human Dipeptidyl-peptidase 1 (heavy chain, Cleaved-Leu231)
Specificity	This antibody detects endogenous levels of Human Dipeptidyl-peptidase 2 (heavy chain, Cleaved-Leu231, protein was cleaved amino acid sequence between 230-231)
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Storage	Store at -20℃ . Avoid repeated freeze-thaw cycles.
Protein Name	Dipeptidyl-peptidase 1 (heavy chain, Cleaved-Leu231)
Gene Name	CTSC CPPI
Cellular localization	Lysosome.
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
Clonality	Polyclonal
Concentration	1 mg/ml
Observed band	18kD
Human Gene ID	1075
Human Swiss-Prot Number	P53634
Alternative Names	Dipeptidyl peptidase 1 (EC 3.4.14.1;Cathepsin C;Cathepsin J;Dipeptidyl peptidase I;DPP-I;DPPI;Dipeptidyl transferase) [Cleaved into: Dipeptidyl peptidase 1 exclusion domain chain





Background

(Dipeptidyl peptidase I exclusion domain chain);
Dipeptidyl peptidase 1 heavy
catalytic activity:Release of an N-terminal dipeptide,
Xaa-Yaa-|-Zaa-, except when Xaa is Arg or Lys, or Yaa
or Zaa is Pro.,cofactor:Binds 1 chloride ion per heavy
chain.,disease:Defects in CTSC are a cause of
Haim-Munk syndrome (HMS) [MIM:245010]; also
known as keratosis palmoplantaris with
periodontopathia and onychogryposis or Cochin
Jewish disorder. HMS is an autosomal recessive
disorder characterized by palmoplantar keratosis,
onychogryphosis and periodontitis. Additional
features are pes planus, arachnodactyly, and
acroosteolysis.,disease:Defects in CTSC are a cause
of juvenile periodontitis (JPD) [MIM:170650]; also
known as prepubertal periodontitis (PPP). JPD is
characterized by severe and protracted gingival
infections, leading to tooth loss. JPD inheritance is
autosomal dominant.,disease:Defects in CTSC are a
cause of Papillon-Lefevre syndrome (PLS)
[MIM:245000]; also known as keratosis
palmoplantaris with periodontopathia. PLS is 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.,enzyme regulation:Strongly inhibited by the
cysteine peptidase inhibitors mersalyl acid,
iodoacetic acid and cystatin. Inhibited by
N-ethylmaleimide, Gly-Phe-diazomethane, TLCK,
TPCK and, at low pH, by dithiodipyridine. Not
inhibited by the serine peptidase inhibitor PMSF, the
aminopeptidase inhibitor bestatin, or metal ion
chelators.,function:Thiol protease. Has
dipeptidylpeptidase activity. Active against a broad
range of dipeptide substrates composed of both
polar and hydrophobic amino acids. Proline cannot
occupy the P1 position and arginine cannot occupy
the P2 position of the substrate. Can act as both an





exopeptidase and endopeptidase. Activates serine proteases such as elastase, cathepsin G and granzymes A and B. Can also activate neuraminidase and factor XIII.,induction:Up-regulated in lymphocytes by IL2.,online information:CTSC mutation db,PTM: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.,PTM:N-glycosylated.,similarity:Belongs to the peptidase C1 family.,subunit:Tetramer of heterotrimers consisting of exclusion domain, heavy- and light chains.,tissue specificity:Ubiquitous. Highly expressed in lung, kidney and placenta. Detected at intermediate levels in colon, small intestine, spleen and pancreas.,

