

Product name:	Cleaved-COL4A3 (L1425) Rabbit Polyclonal Antibody
Cat number:	ABN08984
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human Collagen IV alpha3. AA range:1376-1425
Reactivity:	Human,Rat,Mouse
Applications:	WB 1:500-1:2000,ELISA 1:5000-1:10000
Molecular Weight:	140kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

Type IV collagen, the major structural component of basement membranes, is a multimeric protein composed of 3 alpha subunits. These subunits are encoded by 6 different genes, alpha 1 through alpha 6, each of which can form a triple helix structure with 2 other subunits to form type IV collagen. This gene encodes alpha 3. In the Goodpasture syndrome, autoantibodies bind to the collagen molecules in the basement membranes of alveoli and glomeruli. The epitopes that elicit these autoantibodies are localized largely to the non-collagenous C-terminal domain of the protein. A specific kinase phosphorylates amino acids in this same C-terminal region and the expression of this kinase is upregulated during pathogenesis. This gene is also linked to an autosomal recessive form of Alport syndrome. The mutations contributing to this syndrome are also located within the exons that encode this C-terminal alternative products: The majority of isoforms differ in the C-terminal part of the NC1 domain, disease: Autoantibodies against the NC1 domain of alpha 3(IV) are found in Goodpasture syndrome, an autoimmune disease of lung and kidney, disease: Defects in COL4A3 are a cause of Alport syndrome autosomal recessive (APSAR) [MIM:203780]. APSAR is characterized by progressive glomerulonephritis, glomerular basement membrane defects, renal failure, sensorineural deafness and specific eye abnormalities (lenticonous and macular flecks). The disorder shows considerable heterogeneity in that families differ in the age of end-stage renal disease and the occurrence of deafness, disease: Defects in COL4A3 are a cause of benign familial hematuria (BFH) [MIM:141200]; also known as thin basement membrane nephropathy. BFH is characterized by persistent hematuria, an electron microscopically detectable thin glomerular basement membrane (GBM) and an autosomal dominant mode of inheritance. Renal function remains normal. In children, differentiation between BFH and AS can be difficult, because both disorders are manifested by persistent hematuria and thin GBM at that age, domain: Alpha chains of type IV collagen have a non-collagenous domain (NC1) at their C-terminus, frequent interruptions of the G-X-Y repeats in the long central triple-helical domain (which may cause flexibility in the triple helix), and a short N-terminal triple-helical 7S domain, function: Tumstatin, a cleavage fragment corresponding to the collagen alpha 3(IV) NC1 domain, possesses both anti-angiogenic and anti-tumor cell activity; these two anti-tumor properties may be regulated via RGD-independent ITGB3-mediated mechanisms, function: Type IV collagen is the major structural component of glomerular basement membranes (GBM), forming a 'chicken-wire' meshwork together with laminins, proteoglycans and entactin/nidogen, miscellaneous: The epitopes recognized by the Goodpasture autoantibodies are sequestered within the NC1 hexamer of the type IV collagen network, PTM: Isoform 2 contains an additional N-linked glycosylation site, PTM: Phosphorylated by the Goodpasture antigen-binding protein/COL4A3BP, PTM: Prolines at the third position of the tripeptide repeating unit (G-X-Y) are hydroxylated in some or all of the chains, PTM: Type IV collagens contain numerous cysteine residues which are involved in inter- and intramolecular disulfide bonding. 12 of these, located in the NC1 domain, are conserved in all known type IV collagens, similarity: Belongs to the type IV collagen family, similarity: Contains 1 collagen IV NC1 (C-terminal non-collagenous) domain, subcellular location: Colocalizes with COL4A4 and COL4A5 in GBM, tubular basement membrane (TBM) and synaptic basal lamina (BL), subunit: There are six type IV collagen isoforms, alpha 1(IV)-alpha 6(IV), each of which can form a triple helix structure with 2 other chains to generate type IV collagen network. The alpha 3(IV) chain forms a triple helical protomer with alpha 4(IV) and alpha 5(IV); this triple helical structure dimerizes through NC1-NC1 domain interactions such that the alpha 3(IV), alpha 4(IV) and alpha 5(IV) chains of one protomer connect with the alpha 5(IV), alpha 4(IV) and alpha 3(IV) chains of the opposite protomer, respectively. Interacts with COL4A3BP AND ITGB3. Associates with LAMB2 at the neuromuscular junction and in GBM, tissue specificity: Alpha 3 and alpha 4 type IV collagens are colocalized and present only in basement membranes of kidney, eye, cochlea, lung and brain,.