

Cat. No:	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
Synonyms:	COL4A3; Collagen alpha-3(IV) chain; Goodpasture antigen

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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 (lenticonus 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

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Product Data Sheet:
Cleaved-COL4A3 (L1425) Rabbit Polyclonal Antibody

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