



# Cleaved-COL4A3 (L1425) Monoclonal Antibody

<b>Catalog No</b>	BYmab-16802
<b>Isotype</b>	IgG
<b>Reactivity</b>	Human;Rat;Mouse;
<b>Applications</b>	WB
<b>Gene Name</b>	COL4A3
<b>Protein Name</b>	Collagen alpha-3(IV) chain
<b>Immunogen</b>	The antiserum was produced against synthesized peptide derived from human Collagen IV alpha3. AA range:1376-1425
<b>Specificity</b>	Cleaved-COL4A3 (L1425) Monoclonal Antibody detects endogenous levels of fragment of activated COL4A3 protein resulting from cleavage adjacent to L1425.
<b>Formulation</b>	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
<b>Source</b>	Monoclonal, Mouse,IgG
<b>Purification</b>	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
<b>Dilution</b>	WB 1:500-2000
<b>Concentration</b>	1 mg/ml
<b>Purity</b>	≥90%
<b>Storage Stability</b>	-20°C/1 year
<b>Synonyms</b>	COL4A3; Collagen alpha-3(IV) chain; Goodpasture antigen
<b>Observed Band</b>	140kD
<b>Cell Pathway</b>	Secreted, extracellular space, extracellular matrix, basement membrane. Colocalizes with COL4A4 and COL4A5 in GBM, tubular basement membrane (TBM) and synaptic basal lamina (BL). .
<b>Tissue Specificity</b>	Alpha 3 and alpha 4 type IV collagens are colocalized and present in kidney, eye, basement membranes of lens capsule, cochlea, lung, skeletal muscle, aorta, synaptic fibers, fetal kidney and fetal lung. PubMed:8083201 reports similar levels of expression of alpha 3 and alpha 4 type IV collagens in kidney, but PubMed:7523402 reports that in kidney levels of alpha 3 type IV collagen are significantly lower than those of alpha 4 type IV collagen. According to PubMed:8083201, alpha 3 type IV collagen is not detected in heart, brain, placenta, liver, pancreas, extrasynaptic muscle fibers, endoneurial and perineurial nerves, fetal brain, fetal heart and fetal liver. According to PubMed:7523402, alpha 3 type IV collagen is strongly expressed in pancreas, neuroretina and calvaria and not expressed

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## Function

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 glo

## 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 r

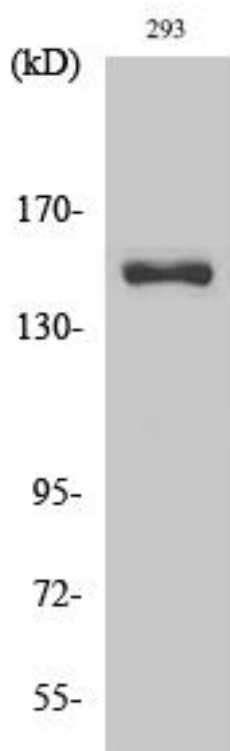
## matters needing attention

Avoid repeated freezing and thawing!

## Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

## Products Images



Western Blot analysis of various cells using Cleaved-COL4A3 (L1425) Monoclonal Antibody