

Product name:	COL4A6 Rabbit Polyclonal Antibody
Cat number:	ABN09190
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 alpha6. AA range:1201-1250
Reactivity:	Human,Rat,Mouse
Applications:	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:200-1:1000,ELISA 1:20000-1:40000
Molecular Weight:	160kDa
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:

This gene encodes one of the six subunits of type IV collagen, the major structural component of basement membranes. Like the other members of the type IV collagen gene family, this gene is organized in a head-to-head conformation with another type IV collagen gene, alpha 5 type IV collagen, so that the gene pair shares a common promoter. Deletions in the alpha 5 gene that extend into the alpha 6 gene result in diffuse leiomyomatosis accompanying the X-linked Alport syndrome caused by the deletion in the alpha 5 gene. Alternative splicing results in multiple transcript variants encoding different isoforms. [provided by RefSeq, Dec 2013],disease:Deletions covering the N-terminal regions of COL4A6 and COL4A5, which are localized in a head-to-head manner, are the cause of diffuse leiomyomatosis with Alport syndrome (DL-ATS) [MIM:308940]; also known as esophageal and vulval leiomyomatosis with nephropathy or Alport syndrome and diffuse leiomyomatosis (ATS-DL). DL-ATS is the combination of Alport syndrome (AS) and diffuse leiomyomatosis (DL). AS is characterized by progressive glomerulonephritis, often associated with high-tone sensorineural deafness, specific eye abnormalities (lenticonus and macular flecks), and glomerular basement membrane defects. DL is a tumorous process involving smooth muscle cells, mostly of the esophagus, but also of the tracheobronchial tree and the female genital tract.,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: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.,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.,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.,