Product Name: COL4A4 Rabbit Polyclonal Antibody
Catalog #: APRab09188
For research use only.

Summary

Description	Rabbit polyclonal Antibody
Host	Rabbit
Application	IHC,ICC/IF,ELISA
Reactivity	Human,Rat,Mouse
Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Concentration	1mg/ml
Storage	Aliquot and store at -20°C (valid for 12 months). Avoid freeze/thaw cycles.
Shipping	Ice bags
Buffer	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
Purification	Affinity purification

Application

Dilution Ratio	IHC 1:100-1:300,ICC/IF 1:200-1:1000,ELISA 1:5000-1:10000
Molecular Weight	

Antigen Information

Gene Name	COL4A4
Alternative Names	COL4A4; Collagen alpha-4(IV) chain
Gene ID	1286.0
SwissProt ID	P53420
Immunogen	The antiserum was produced against synthesized peptide derived from human Collagen IV alpha4. AA range:541-590

Background

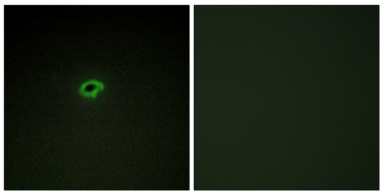
This gene encodes one of the six subunits of type IV collagen, the major structural component of basement membranes. This

particular collagen IV subunit, however, is only found in a subset 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 so that each gene pair shares a common promoter. Mutations in this gene are associated with type II autosomal recessive Alport syndrome (hereditary glomerulonephropathy) and with familial benign hematuria (thin basement membrane disease). Two transcripts, differing only in their transcription start sites, have been identified for this gene and, as is common for collagen genes, multiple polyadenylation sites are found in the 3' UTR. [provided by RefSeq, Jul 2008],disease:Defects in COL4A4 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 COL4A4 are a cause of benign familial hematuria (BFH) [MIM:141200]; also known as thin basement membrane disease. 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: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.,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. 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.,

Research Area

Focal adhesion;ECM-receptor interaction;Pathways in cancer;Small cell lung cancer;

Image Data



Immunofluorescence analysis of COS7 cells, using Collagen IV alpha4 Antibody. The picture on the right is blocked with the synthesized peptide.