abcam

Product datasheet

Anti-COL4A3 antibody ab85103

1 图像

概述

产品名称	Anti-COL4A3抗体
描述	**兔多克隆抗体**to COL4A3
宿主	Rabbit
经测试应用	**适用于:** IHC-P
种属反应性	**与反应:** Human
免疫原	Synthetic peptide (Human) from the N-terminal of COL4A3
阳性对照	Human kidney

性能

形式	Lyophilised:Reconstitute in 200ul sterile water.
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
存储溶液	Preservative: 0.02% Sodium Azide Constituents: 2% BSA, 0.1M Tris glycine, pH 7.2
纯度	Immunogen affinity purified
克隆	**多克隆**
同种型	IgG

应用

Our Abpromise guarantee covers the use of **ab85103** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

应用	Ab评论	说明
IHC-P		1/50 - 1/200.

靶标

功能	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. 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.

组织特异性

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 in adrenal, ileum and skin. Isoform 1 and isoform 3 are strongly expressed in kidney, lung, suprarenal capsule, muscle and spleen, in each of these tissues isoform 1 is more abundant than isoform 3. Isoform 1 and isoform 3 are expressed at low levels in artery, fat, pericardium and peripherical nerve, but not in placenta, mesangium, skin, pleura and cultured umbilical endothelial cells.

疾病相关

Note=Autoantibodies against the NC1 domain of alpha 3(IV) are found in Goodpasture syndrome, an autoimmune disease of lung and kidney.
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.
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.
Defects in COL4A3 are a cause of Alport syndrome autosomal dominant (APSAD) [MIM:104200]. Alport syndrome 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.

序列相似性

Belongs to the type IV collagen family.
Contains 1 collagen IV NC1 (C-terminal non-collagenous) domain.

结构域

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.

翻译后修饰

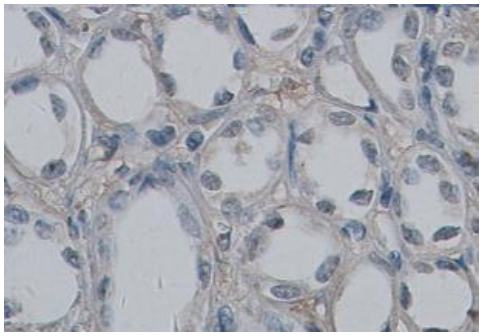
Prolines at the third position of the tripeptide repeating unit (G-X-Y) are hydroxylated in some or all of the chains.
Isoform 2 contains an additional N-linked glycosylation site.
Type IV collagens contain numerous cysteine residues which are involved in inter- and intramolecular disulfide bonding. 12 of these, located in the NC1 domain, are conserved in all known type IV collagens.
The trimeric structure of the NC1 domains is stabilized by covalent bonds between Lys and Met residues.
Phosphorylated by the Goodpasture antigen-binding protein/COL4A3BP.

细胞定位

Secreted > extracellular space > extracellular matrix > basement membrane. Colocalizes with

COL4A4 and COL4A5 in GBM, tubular basement membrane (TBM) and synaptic basal lamina (BL).

图片



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-COL4A3 antibody (ab85103)

ab85103 at 1/100 dilution staining COL4A3 in formalin-fixed, paraffin-embedded human kidney cells by immunohistochemistry.

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit http://www.abcam.cn/abpromise or contact our technical team.

Terms and conditions

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors