# abcam

# Product datasheet

# Recombinant Human COL4A3 protein (His tag) ab275563

# 1 Image

**Description** 

Product name Recombinant Human COL4A3 protein (His tag)

Purity > 97 % SDS-PAGE.

Expression system < 1.000 Eu/µg
Expression system

Accession Q01955

Protein length Protein fragment

Animal free No

Nature Recombinant

**Species** Human

**Sequence** LKG KRGDSGSPAT WTTRGFVFTR HSQTTAIPSC

PEGTVPLYSG FSFLFVQGNQ RAHGQDLGTL GSCLQRFTTM PFLFCNVNDV CNFASRNDYS

YWLSTPALMP MNMAPITGRA LEPYISRCTV CEGPAIAIAV

HSQTTDIPPC PHGWISLWKG FSFIMFTSAG SEGTGQALAS PGSCLEEFRA SPFLECHGRG

TCNYYSNSYS FWLASLNPER MFRKPIPSTV KAGELEKIIS

**RCQVCMKKRH** 

Predicted molecular weight 31 kDa

**Molecular weight information** 30kDa by SDS-PAGE reducing conditions.

Amino acids 1428 to 1670

Tags His tag N-Terminus

## **Specifications**

Our **Abpromise guarantee** covers the use of **ab275563** in the following tested applications.

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

Applications SDS-PAGE

Form Lyophilized

1

#### **Preparation and Storage**

### Stability and Storage

Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -80°C.

Avoid freeze / thaw cycle.

pH: 8.00

Constituents: 16.38% PBS, 0.17% Sodium-N-Lauroylsarcosinate, 82.8% Trehalose

#### Reconstitution

Reconstitute in 20mM Tris, 150mM NaCl (pH8.0) to a concentration of 0.1-1.0 mg/mL. Do not

vortex.

#### **General Info**

#### **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. 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.

#### Tissue specificity

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.

#### Involvement in disease

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.

# Sequence similarities

Belongs to the type IV collagen family.

Contains 1 collagen IV NC1 (C-terminal non-collagenous) domain.

**Domain** Alpha chains of type V 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.

Post-translational modifications

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.

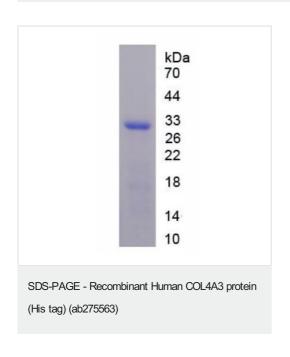
Phosphorylated by the Goodpasture antigen-binding protein/COL4A3BP.

**Cellular localization** Secreted > extracellular space > extracellular matrix > basement membrane. Colocalizes with

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

(BL).

### **Images**



SDS-PAGE analysis of ab275563.

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