



## Rabbit Anti-procollagen type IIA antibody

SL11069R

<b>Product Name:</b>	procollagen type IIA
<b>Chinese Name:</b>	II 型胶原 $\alpha$ 1 N端前体肽抗体
<b>Alias:</b>	Collagen II N-terminal propeptide; Collagen II alpha 1; COL2A1; COL2A1 protein; collagen, type II, alpha 1; collagen alpha-1(II); type II collagen; alpha-1 type II collagen; alpha1 type II collagen; Col2a1; AOM; Cartilage collagen; COL11A3; Collagen alpha 1(II) chain precursor; Collagen II alpha 1 polypeptide; Collagen type II alpha 1 (primary osteoarthritis spondyloepiphyseal dysplasia congenital); MGC131516; SEDC; Collagen alpha-1(II) chain; Alpha-1 type II collagen; CO2A1 HUMAN.
<b>Organism Species:</b>	Rabbit
<b>Clonality:</b>	Polyclonal
<b>React Species:</b>	Human,Chicken,Dog,Pig,Cow,Rabbit,Sheep,Guinea Pig,Danio rerio
<b>Applications:</b>	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
<b>Molecular weight:</b>	17/163kDa
<b>Cellular localization:</b>	Extracellular matrixSecretory protein
<b>Form:</b>	Lyophilized or Liquid
<b>Concentration:</b>	1mg/ml
<b>immunogen:</b>	KLH conjugated synthetic peptide derived from human Collagen II alpha 1 N-terminal propeptide:1-100/1487
<b>Lsotype:</b>	IgG
<b>Purification:</b>	affinity purified by Protein A
<b>Storage Buffer:</b>	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
<b>Storage:</b>	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
<b>PubMed:</b>	<a href="#">PubMed</a>
<b>Product Detail:</b>	Collagen type II is the most abundant protein of cartilage matrix. Two isoforms exist

and collagen type IIA is involved in early development of cartilage in embryos. The collagen is produced from type II procollagen when two teloterminal propeptides are digested from the precursor and released into the circulation. Procollagen type II N-terminal propeptide is considered a marker of cartilage synthesis and the normal concentration in serum is approximately 30ng/mL.

**Function:**

Type II collagen is specific for cartilaginous tissues. It is essential for the normal embryonic development of the skeleton, for linear growth and for the ability of cartilage to resist compressive forces.

**Subunit:**

Homotrimers of alpha 1(II) chains.

**Subcellular Location:**

Secreted, extracellular space, extracellular matrix.

**Tissue Specificity:**

Isoform 2 is highly expressed in juvenile chondrocyte and low in fetal chondrocyte.

**Post-translational modifications:**

Probably 3-hydroxylated on prolines by LEPREL1 (By similarity). Proline residues at the third position of the tripeptide repeating unit (G-X-P) are hydroxylated in some or all of the chains. Proline residues at the second position of the tripeptide repeating unit (G-P-X) are hydroxylated in some of the chains.

The N-telopeptide is covalently linked to the helical COL2 region of alpha 1(IX), alpha 2(IX) and alpha 3(IX) chain. The C-telopeptide is covalently linked to another site in the helical region of alpha 3(IX) COL2.

**DISEASE:**

Defects in COL2A1 are the cause of spondyloepiphyseal dysplasia congenital type (SEDC) [MIM:183900]. This disorder is characterized by disproportionate short stature and pleiotropic involvement of the skeletal and ocular systems.

Defects in COL2A1 are the cause of spondyloepimetaphyseal dysplasia Strudwick type (SEMD-STR) [MIM:184250]. A bone disease characterized by disproportionate short stature from birth, with a very short trunk and shortened limbs, and skeletal abnormalities including lordosis, scoliosis, flattened vertebrae, pectus carinatum, coxa vara, clubfoot, and abnormal epiphyses or metaphyses. A distinctive radiographic feature is irregular sclerotic changes, described as dappled in the metaphyses of the long bones.

Defects in COL2A1 are the cause of achondrogenesis type 2 (ACG2) [MIM:200610]; also known as achondrogenesis-hypochondrogenesis type II. ACG2 is a disease characterized by the absence of ossification in the vertebral column, sacrum and pubic bones.

Defects in COL2A1 are the cause of Legg-Calve-Perthes disease (LCPD) [MIM:150600]; also known as Legg-Perthes disease or Perthes disease. LCPD is

characterized by loss of circulation to the femoral head, resulting in avascular necrosis in a growing child. Clinical pictures of the disease vary, depending on the phase of disease progression through ischemia, revascularization, fracture and collapse, and repair and remodeling of the bone.

Defects in COL2A1 are the cause of Kniest dysplasia (KD) [MIM:156550]; also known as Kniest syndrome or metatropic dwarfism type II. KD is a moderately severe chondrodysplasia phenotype that results from mutations in the COL2A1 gene. Characteristics of the disorder include a short trunk and extremities, mid-face hypoplasia, cleft palate, myopia, retinal detachment, and hearing loss.

Defects in COL2A1 are a cause of primary avascular necrosis of femoral head (ANFH) [MIM:608805]; also known as ischemic necrosis of the femoral head or osteonecrosis of the femoral head. ANFH causes disability that often requires surgical intervention. Most cases are sporadic, but families in which there is an autosomal dominant inheritance of the disease have been identified. It has been estimated that 300,000 to 600,000 people in the United States have ANFH. Approximately 15,000 new cases of this common and disabling disorder are reported annually. The age at the onset is earlier than that for osteoarthritis. The diagnosis is typically made when patients are between the ages of 30 and 60 years. The clinical manifestations, such as pain on exertion, a limping gait, and a discrepancy in leg length, cause considerable disability. Moreover, nearly 10 percent of the 500,000 total-hip arthroplasties performed each year in the United States involve patients with ANFH. As a result, this disease creates a substantial socioeconomic cost as well as a burden for patients and their families.

Defects in COL2A1 are the cause of osteoarthritis with mild chondrodysplasia (OACD) [MIM:604864]. Osteoarthritis is a common disease that produces joint pain and stiffness together with radiologic evidence of progressive degeneration of joint cartilage. Some forms of osteoarthritis are secondary to events such as trauma, infections, metabolic disorders, or congenital or heritable conditions that deform the epiphyses or related structures. In most patients, however, there is no readily identifiable cause of osteoarthritis. Inheritance in a Mendelian dominant manner has been demonstrated in some families with primary generalized osteoarthritis. Reports demonstrate coinheritance of primary generalized osteoarthritis with specific alleles of the gene COL2A1, the precursor of the major protein of cartilage.

Defects in COL2A1 are the cause of platyspondylic lethal skeletal dysplasia Torrance type (PLSD-T) [MIM:151210]. Platyspondylic lethal skeletal dysplasias (PLSDs) are a heterogeneous group of chondrodysplasias characterized by severe platyspondyly and limb shortening. PLSD-T is characterized by varying platyspondyly, short ribs with anterior cupping, hypoplasia of the lower ilia with broad ischial and pubic bones, and shortening of the tubular bones with splayed and cupped metaphyses. Histology of the growth plate typically shows focal hypercellularity with slightly enlarged chondrocytes in the resting cartilage and relatively well-preserved columnar formation and ossification at the chondro-osseous junction. PLSD-T is generally a perinatally lethal disease, but a few long-term survivors have been reported.

Defects in COL2A1 are the cause of multiple epiphyseal dysplasia with myopia and conductive deafness (EDMMD) [MIM:132450]. Multiple epiphyseal dysplasia is a generalized skeletal dysplasia associated with significant morbidity. Joint pain, joint deformity, waddling gait, and short stature are the main clinical signs and symptoms.

EDMMD is an autosomal dominant disorder characterized by epiphyseal dysplasia associated with progressive myopia, retinal thinning, crenated cataracts, conductive deafness.

Defects in COL2A1 are the cause of spondyloperipheral dysplasia (SPD) [MIM:271700]. SPD patients manifest short stature, midface hypoplasia, sensorineural hearing loss, spondyloepiphyseal dysplasia, platyspondyly and brachydactyly.

**Similarity:**

Belongs to the fibrillar collagen family.  
Contains 1 fibrillar collagen NC1 domain.  
Contains 1 VWFC domain.

**SWISS:**

P02458

**Gene ID:**

1280

**Database links:**

[Entrez Gene: 1280](#)Human

[Omim: 120140](#)Human

[SwissProt: P02458](#)Human

[Unigene: 408182](#)Human

**Important Note:**

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.