

Collagen II Rabbit pAb

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Catalog # AP94387

Product Information

Application	WB
Reactivity	Rabbit
Host	Rabbit
Clonality	Polyclonal
Calculated MW	117 KDa
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from rabbit Collagen II Chondrocalcin
Epitope Specificity	1401-1487/1487
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Secreted, extracellular space, extracellular matrix.
SIMILARITY	Belongs to the fibrillar collagen family.Contains 1 fibrillar collagen NC1 domain.Contains 1 VWFC domain.
SUBUNIT	Homotrimers of alpha 1(II) chains.
Post-translational modifications	Proline residues at the third position of the tripeptide repeating unit (G-X-Y) are hydroxylated in some or all of the chains. Proline residues at the second position of the tripeptide repeating unit (G-X-Y) 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 an 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.

Important Note

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

Background Descriptions

This gene encodes the alpha-1 chain of type II collagen, a fibrillar collagen found in cartilage and the vitreous humor of the eye. Mutations in this gene are associated with achondrogenesis, chondrodysplasia, early onset familial osteoarthritis, SED congenita, Langer-Saldino achondrogenesis, Kniest dysplasia, Stickler syndrome type I, and spondyloepimetaphyseal dysplasia Strudwick type. In addition, defects in processing chondrocalcin, a calcium binding protein that is the C-propeptide of this collagen molecule, are also

associated with chondrodysplasia. There are two transcripts identified for this gene. [provided by RefSeq, Jul 2008]

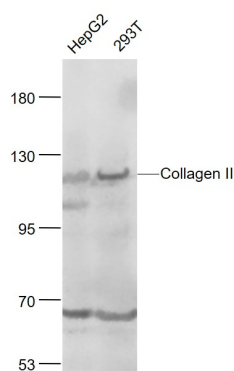
Additional Information

Target/Specificity	Isoform 2 is highly expressed in juvenile chondrocyte and low in fetal chondrocyte.
Dilution	WB=1:500-2000
Format	0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

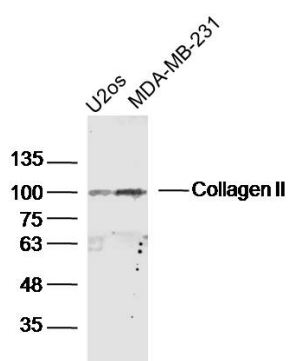
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Images



Sample: HepG2(Human) Cell Lysate at 30 ug
293T(Human) Cell Lysate at 30 ug Primary: Anti- Collagen II (AP94387) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 116 kD Observed band size: 116 kD



Sample: U2OS Cell (Human) Lysate at 40 ug MDA-MB-231 Cell (Human) Lysate at 40 ug Primary: Anti-Collagen II (AP94387) at 1/300 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 117/163 kD Observed band size: 100 kD

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