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WISP3 Rabbit pAb

Catalog # AP98853

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	O95389
Other Accession	O95389
Reactivity	Rat
Predicted	Human, Mouse, Rabbit, Monkey, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	39293
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human WISP3
Epitope Specificity	221-320/354
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the CCN family. Contains 1 CTCK (C-terminal cystine knot-like) domain. Contains 1 IGFBP N-terminal domain. Contains 1 TSP type-1 domain.
DISEASE	Defects in WISP3 are the cause of progressive pseudorheumatoid arthropathy of childhood (PPAC) [MIM:208230]. PPAC is an autosomal recessive disorder characterized by stiffness and swelling of joints, motor weakness and joint contractures. Signs and symptoms of the disease develop typically between three and eight years of age. This progressive disease is a primary disorder of articular cartilage with continued cartilage loss and destructive bone changes with aging.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Wnt-induced secreted protein (WISP)-1, WISP-2 and WISP-3 are members of the CCN family of growth factors, which include connective tissue growth factor (CTGF) and Cyr61. WISP-1, WISP-2 and WISP-3 share significant sequence similarity, including four conserved cysteine-rich domains, and they are believed to function as dimers in their active forms. WISP-1 expression is observed in various tissues including adult heart, kidney and spleen, while WISP-2 expression predominates in skeletal muscle, colon and ovary. Both WISP-1 and WISP-2 are upregulated in cells transformed with the proto-oncogene Wnt-1, and they are also more highly expressed in human colon tumors, suggesting that these proteins may participate in tumor development. WISP-3 is involved in normal post-natal skeletal growth, and it is also implicated in the development of the autosomal recessive skeletal disorder progressive pseudorheumatoid dysplasia, which affects cartilage homeostasis by disrupting the growth of chondrocyte and normal cell columnar organization.

Additional Information

Gene ID	8838
Other Names	RGD1564120; Wisp3; CCN6_HUMAN; CCN6; CCN family member 6; WNT1-inducible-signaling pathway protein 3 (WISP-3); CCN6_MOUSE; cellular communication network factor 6; WNT1 inducible signaling pathway protein 3
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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.

Protein Information

Name	CCN6 (HGNC:12771)
Function	Plays a role in mitochondrial electron transport and mitochondrial respiration (PubMed:27252383). Through its regulation of the mitochondrial function may play a role in normal postnatal skeletal growth and cartilage homeostasis (PubMed:10471507, PubMed:27252383).
Cellular Location	Secreted. Mitochondrion. Note=Associated with membranes.
Tissue Location	Predominant expression in adult kidney and testis and fetal kidney. Weaker expression found in placenta, ovary, prostate and small intestine (PubMed:10471507, PubMed:9843955). Also expressed in skeletally-derived cells such as synoviocytes and articular cartilage chondrocytes (PubMed:10471507).

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