

Sclerostin Rabbit pAb

Catalog # AP99923

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q9BQB4
Other Accession	Q9BQB4
Host	Rabbit
Clonality	Polyclonal
Calculated MW	24031
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Sclerostin
Epitope Specificity	151-213/213
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 sclerostin family. Contains 1 CTCK (C-terminal cystine knot-like) domain.
SUBUNIT	Interacts with LRP4 (via the extracellular domain); the interaction facilitates the inhibition of Wnt signaling. Interacts with LRP5 (via the first two YWTD-EGF repeat domains); the interaction inhibits Wnt-mediated signaling. Interacts with LRP6.
DISEASE	Defects in SOST are the cause of sclerosteosis (SOST) [MIM:269500]; also known as cortical hyperostosis with syndactyly. SOST is an autosomal recessive sclerosing bone dysplasia characterized by a generalized hyperostosis and sclerosis leading to a markedly thickened skull, with mandible, ribs, clavicles and all long bones also being affected. Due to narrowing of the foramina of the cranial nerves, facial nerve palsy, hearing loss and atrophy of the optic nerves can occur. Sclerosteosis is clinically and radiologically very similar to van Buchem disease, mainly differentiated by hand malformations and a large stature in sclerosteosis patients. Note=A 52 kb deletion downstream of SOST results in SOST transcription suppression and is a cause of van Buchem disease (VBCH) [MIM:239100]; also known as hyperostosis corticalis generalisata. VBCH is an autosomal recessive sclerosing bone dysplasia characterized by endosteal hyperostosis of the mandible, skull, ribs, clavicles, and diaphyses of the long bones. Affected patients present a symmetrically increased thickness of bones, most frequently found as an enlarged jawbone, but also an enlargement of the skull, ribs, diaphysis of long bones, as well as tubular bones of hands and feet. The clinical consequence of increased thickness of the skull include facial nerve palsy causing hearing loss, visual problems, neurological pain, and, very rarely, blindness as a consequence of optic atrophy. Serum alkaline phosphatase levels are elevated.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Negative regulator of bone growth.Sclerostin (SOST) is a bone morphogenetic protein (BMP) antagonist, leading to the activation of BMP signaling. It negatively regulates the formation of bone by repressing the differentiation and/or function of osteoblasts induced by BMPs. It has been shown that

Sclerostin binds BMP-5, -6, and -7 with high affinity and BMP-2 and -4 with low affinity. The noggin-sclerostin protein complex represents a novel mechanism for the fine-tuning of BMP activity in bone homeostasis. Evidence is accumulating that one of the important mechanisms of bone regulation by sclerostin is the modulation of Wnt/Beta-catenin signaling. Sclerostin also rapidly activated ERK-1/2 MAPK signaling, indicating the involvement of additional signaling pathways

Additional Information

Gene ID	50964
Other Names	CDD; DAND6; SOST1; VBCH; 5430411E23Rik; SOST_HUMAN; SOST; SOST_MOUSE; SOST_RAT; sclerostin; sclerosteosis
Dilution	WB=1:500-2000,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	SOST (HGNC:13771)
Function	Negative regulator of bone growth that acts through inhibition of Wnt signaling and bone formation.
Cellular Location	Secreted, extracellular space, extracellular matrix
Tissue Location	Widely expressed at low levels with highest levels in bone, cartilage, kidney, liver, bone marrow and primary osteoblasts differentiated for 21 days. Detected in the subendothelial layer of the aortic intima (at protein level).

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.