

Rabbit Anti-Sclerostin antibody

SL10200R

Product Name:	Sclerostin
Chinese Name:	骨形态发生抑制蛋白SOST抗体
Alias:	BEER; Cortical hyperostosis with syndactyly; Sclerosteosis; Sclerostin; SOST; SOST_HUMAN; VBCH.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human, Mouse, Rat, Dog, Pig, Cow, Horse, Rabbit, Sheep,
Applications:	WB=1:500-2000ELISA=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.
Molecular weight:	21kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Sclerostin:121-180/213
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	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.
PubMed:	PubMed
Product Detail:	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

Function:

Negative regulator of bone growth that acts through inhibition of Wnt signaling and bone formation.

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.

Subcellular Location:

Secreted.

Tissue Specificity:

Widely expressed at low levels with highest levels in bone, cartilage, kidney, liver, bone marrow and primary osteeoblasts differentiated for 21 days.

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.

Similarity:

Belongs to the sclerostin family. Contains 1 CTCK (C-terminal cystine knot-like) domain.

SWISS: Q9BQB4 Gene ID: 50964 Database links: Entrez Gene: 50964Human Entrez Gene: 74499Mouse jotech. on Entrez Gene: 80722Rat Omim: 605740Human SwissProt: Q9BQB4Human SwissProt: Q99P68Mouse SwissProt: Q99P67Rat Unigene: 349204Human Unigene: 265602Mouse Unigene: 95369Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.



