



Shh Rabbit mAb

Catalog No	YP-rAb-17317
Isotype	IgG
Reactivity	Human,Mouse,Rat
Applications	WB,IHC,IF,ELISA
Gene Name	SHH
Protein Name	Shh
Purification Process	Protein A
Specificity	Endogenous
Formulation	PBS, 50% glycerol, 0.05% Proclin 300, 0.05%BSA
Source	Monoclonal, Rabbit,IgG
Dilution	IHC 1:200-1:1000; WB 1:2000-1:10000; IF 1:200-1:1000; ELISA 1:5000-1:20000; Note: For IHC, we suggest antigen retrieval with TE buffer pH 9.0
Concentration	0.5 mg/ml
Purity	≥90%
Storage Stability	-15 ° C to -25 ° C/1 year(Do not lower than -25 ° C)
Synonyms	Shh;Sonic hedgehog protein ; SHH ; HHG-1 ; [Cleaved into: Sonic hedgehog protein N-product ; Sonic hedgehog protein C-product]
Observed Band	27kD
Calculated Molecular Weight	50kD
Cell Pathway	Endoplasmic reticulum membrane . Golgi apparatus membrane . Co-localizes with HHAT in the ER and Golgi membrane. . ; [Sonic hedgehog protein N-product]: Cell membrane ; Lipid-anchor . The dual-lipidated sonic hedgehog protein N-product (ShhNp) is firmly tethered to the cell membrane where it forms multimers (PubMed:24522195). Further solubilization and release from the cell surface seem to be achieved through different mechanisms, including the interaction with DISP1 and SCUBE2, movement by lipoprotein particles, transport by cellular extensions called cytonemes or by the proteolytic removal of both terminal lipidated peptides (PubMed:26875496, PubMed:24522195). .
Tissue Specificity	Fetal lung,Plasma,
Function	Disease:Defects in SHH are a cause of solitary median maxillary central incisor (SMMCI) [MIM:147250]. SMMCI is a rare dental anomaly characterized by the congenital absence of one maxillary central incisor..Disease:Defects in SHH are the cause of holoprosencephaly type 3 (HPE3) [MIM:142945]. Holoprosencephaly (HPE) [MIM:236100] is the most common structural anomaly of the brain, in which





the developing forebrain fails to correctly separate into right and left hemispheres. Holoprosencephaly is genetically heterogeneous and associated with several distinct facies and phenotypic variability. The majority of HPE3 cases are apparently sporadic, although clear examples of autosomal dominant inheritance have been described. Interestingly, up to 30% of obligate carriers of HPE3 gene in autosomal dominant pedigrees are clinically unaffected. Disease: Defects in SHH are the cause of microphthalmia isolated with coloboma type 5 (MCOPCB5) [MIM:611638]. Microphthalmia is a clinically heterogeneous disorder of eye formation, ranging from small size of a single eye to complete bilateral absence of ocular tissues. Ocular abnormalities like opacities of the cornea and lens, scarring of the retina and choroid, cataract and other abnormalities like cataract may also be present. Ocular colobomas are a set of malformations resulting from abnormal morphogenesis of the optic cup and stalk, and the fusion of the fetal fissure (optic fissure). Disease: Defects in SHH are the cause of triphalangeal thumb-polysyndactyly syndrome (TPTPS) [MIM:174500]. TPTPS is an autosomal dominant syndrome characterized by a wide spectrum of pre- and post-axial abnormalities due to altered SHH expression pattern during limb development. TPTPS mutations have been mapped to the 7q36 locus in the LMBR1 gene which contains in its intron 5 a long-range cis-regulatory element of SHH expression. Function: Binds to the patched receptor, which functions in association with smoothened (SMO), to activate the transcription of target genes. In the absence of SHH, PTC represses the constitutive signaling activity of SMO. Also regulates another target, the gli oncogene. Intercellular signal essential for a variety of patterning events during development: signal produced by the notochord that induces ventral cell fate in the neural tube and somites, and the polarizing signal for patterning of the anterior-posterior axis of the developing limb bud. Displays both floor plate- and motor neuron-inducing activity. The threshold concentration of N-product required for motor neuron induction is 5-fold lower than that required for floor plate induction. mass spectrometry: Membrane-bound N-product, purified from insect cells PubMed:9593755, mass spectrometry: Soluble N-product, purified from insect cells PubMed:9593755, PTM: Cholesterylolation is required for N-product targeting to lipid rafts and multimerization. PTM: N-palmitoylation of Cys-24 by HHAT is required for N-product multimerization and full activity. PTM: The C-terminal domain displays an autoproteolysis activity and a cholesterol transferase activity. Both activities result in the cleavage of the full-length protein and covalent attachment of a cholesterol moiety to the C-terminal of the newly generated N-terminal fragment (N-product). The N-product is the active species in both local and long-range signaling, whereas the C-product has no signaling activity. similarity: Belongs to the hedgehog family. subcellular location: The C-terminal peptide diffuses from the cell. subcellular location: The N-product either remains associated with lipid rafts at the cell surface, or forms freely diffusible active multimers with its hydrophobic lipid-modified N- and C-termini buried inside. subunit: Interacts with HHATL/GUP1 which negatively regulates HHAT-mediated palmitoylation of the SHH N-terminus. N-product is active as a multimer. tissue specificity: Expressed in fetal intestine, liver, lung, and kidney. Not expressed in adult tissues.

Background

This gene encodes a protein that is instrumental in patterning the early embryo. It has been implicated as the key inductive signal in patterning of the ventral neural tube, the anterior-posterior limb axis, and the ventral somites. Of three human proteins showing sequence and functional similarity to the sonic hedgehog protein of Drosophila, this protein is the most similar. The protein is made as a precursor that is autocatalytically cleaved; the N-terminal portion is soluble and contains the signalling activity while the C-terminal portion is involved in precursor processing. More importantly, the C-terminal product covalently attaches a cholesterol moiety to the N-terminal product, restricting the N-terminal product to the cell surface and preventing it from freely diffusing throughout the developing embryo. Defects in this protein or in its signalling pathway are a cause of holoprosencephaly (HPE), a

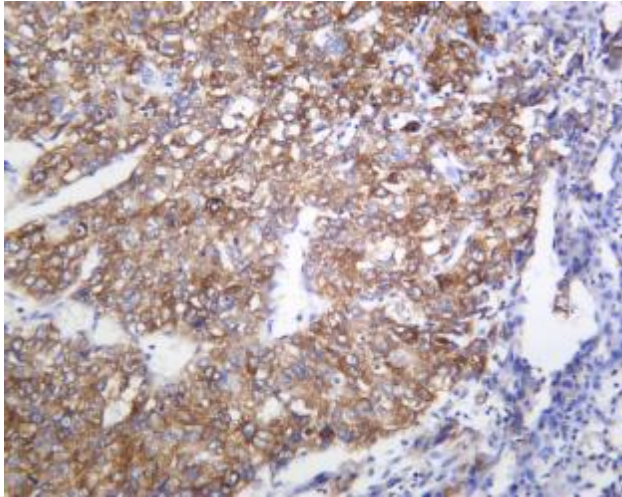
matters needing attention

Avoid repeated freezing and thawing!

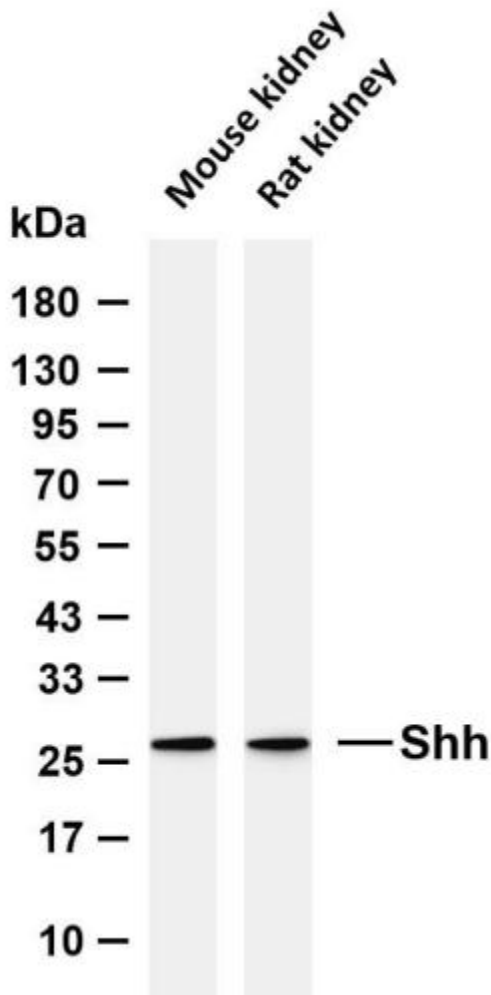
Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.





Human hepatocellular carcinoma was stained with anti-Shh Rabbit antibody



Various whole cell lysates were separated by 4-20% SDS-PAGE, and the membrane was blotted with anti-Shh antibody. The HRP-conjugated Goat anti-Rabbit IgG (H + L) antibody was used to detect the antibody. Lane 1: Mouse kidney Lane 2: Rat kidney Predicted band size: 50kDa Observed band size: 27kDa

