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Shh Rabbit pAb

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Catalog # AP52268

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q15465
Reactivity	Rat, Mouse
Predicted	Human, Rabbit, Chicken, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	49607
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Shh
Epitope Specificity	21-120/462
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	Sonic hedgehog protein C-product: Secreted, extracellular space. Note=The C-terminal peptide diffuses from the cell. Sonic hedgehog protein N-product: Cell membrane; Lipid-anchor. Note=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.
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, scaring 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).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. 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. 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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	SHH binds to the patched (PTC) 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 (By similarity).

Additional Information

Gene ID	6469
Other Names	Sonic hedgehog protein, SHH, 3.1.-.-, HHG-1, Shh unprocessed N-terminal signaling and C-terminal autoprocessing domains, ShhNC, Sonic hedgehog protein N-product, ShhN, Shh N-terminal processed signaling domains, ShhNp, SHH (HGNC:10848)
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	SHH {ECO:0000303|PubMed:7590746, ECO:0000312|HGNC:HGNC:10848}
Function	[Sonic hedgehog protein]: Precursor of sonic hedgehog, a morphogen that activates the smoothened signaling pathway, and which is essential for a variety of patterning events during development (PubMed:29954986, PubMed:29995851, PubMed:30139912, PubMed:31127104, PubMed:31548691). The C-terminal part of the precursor displays an autoproteolysis and a cholesterol transferase activity, resulting (1) in the cleavage of the full-length protein into two parts, Sonic hedgehog protein N-product and C-product (ShhN and ShhC, respectively) and (2) covalent attachment of a cholesterol moiety to the C-terminus of the newly generated ShhN (By similarity). Both autoproteolysis and a cholesterol transferase activities occur in the endoplasmic reticulum (By similarity). Following additional lipidation, ShhN acts as a morphogen, while ShhC is degraded in the endoplasmic reticulum (By similarity).
Cellular Location	[Sonic hedgehog protein]: Endoplasmic reticulum membrane. Golgi apparatus membrane. Note=Co-localizes with HHAT in the ER and Golgi membrane.

Background

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References

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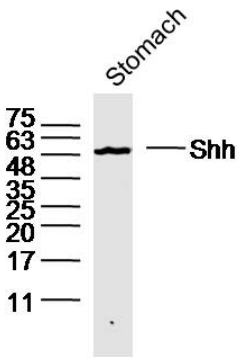
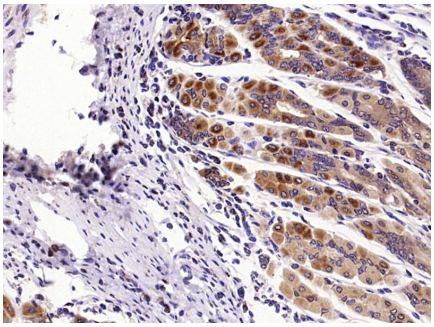
Hillier L.W.,et al.Nature 424:157-164(2003).
Scherer S.W.,et al.Science 300:767-772(2003).
Pepinsky R.B.,et al.J. Biol. Chem. 273:14037-14045(1998).

Images



Sample:Stomach (Mouse) Lysate at 40 ug
Primary: Anti-Shh(AP52268)at 1/300 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 19/47kD
Observed band size: 49 kD



Paraformaldehyde-fixed, paraffin embedded (Mouse stomach); Antigen retrieval by microwave in sodium citrate buffer (pH6.0) ; Block endogenous peroxidase by 3% hydrogen peroxide for 30 minutes; Blocking buffer (3% BSA) at RT for 30min; Antibody incubation with (Shh) Polyclonal Antibody, Unconjugated (AP52268) at 1:400 overnight at 4℃, followed by conjugation to the secondary antibody (labeled with HRP)and DAB staining.