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# Shh Rabbit pAb

CatalogNo: YT6188

# Key Features

40kD (Observed)

Host SpeciesRabbit

MW

ReactivityHuman,Mouse,Rat

Isotype

IgG

ApplicationsIHC,IF,WB

#### **Recommended Dilution Ratios**

IHC 1:50-200 WB 1:500-2000 IF 1:50-200

# **Storage**

Storage\*-15°C to -25°C/1 year(Do not lower than -25°C)FormulationLiquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

# **Basic Information**

Clonality Polyclonal

# Immunogen Information

Immunogen	Synthesized peptide derived from human Shh
Specificity	This antibody detects endogenous levels of human Shh

# **Target Information**

SHH

Gene name

	Organism	Gene ID	UniProt ID
	Human	<u>6469;</u>	<u>Q15465;</u>
Cellular Localization	the ER and Golgi membrane; [S anchor . The dual-lipidated sonic the cell membrane where it forms	tic removal of both terminal	

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 exemples 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, 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).,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 (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., mass spectrometry: Membrane-bound N-product, purified from insect cells PubMed:9593755,mass spectrometry:Soluble N-product, purified from insect cells PubMed:9593755, PTM: Cholesterylation 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 Nterminus. N-product is active as a multimer., tissue specificity: Expressed in fetal intestine, liver, lung, and kidney. Not expressed in adult tissues.,

#### Validation Data

#### **Contact information**

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Please scan the QR code to access additional product information: **Shh Rabbit pAb** 

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