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Immunotag™ Shh Monoclonal Antibody

Antibody Specification

Catalog No.	ITM0577
Product Description	Immunotag™ Shh Monoclonal Antibody
Size	50 µg, 100 µg
Conjugation	HRP, Biotin, FITC, Alexa Fluor® 350, Alexa Fluor® 405, Alexa Fluor® 488, Alexa Fluor® 555, Alexa Fluor® 594, Alexa Fluor® 647
IMPORTANT NOTE	This product is custom manufactured with a lead time of 3-4 weeks. Once in production, this item cannot be cancelled from an order and is not eligible for return.
Target Protein	SHH
Clonality	Monoclonal
Storage/Stability	-20°C/1 year
Application	WB,ELISA
Recommended Dilution	Western Blot: 1/500 - 1/2000. ELISA: 1/10000. Not yet tested in other applications.
Concentration	1 mg/ml
Reactive Species	Human
Host Species	Mouse
Immunogen	Purified recombinant fragment of human Shh expressed in E. Coli.
Specificity	Shh Monoclonal Antibody detects endogenous levels of Shh protein.
Purification	Affinity purification
Form	Ascitic fluid containing 0.03% sodium azide.
Gene Name	SHH
Accession No.	Q15465 Q62226
Alternate Names	SHH; Sonic hedgehog protein; SHH; HHG-1

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Description	sonic hedgehog(SHH) Homo sapiens 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 d
Cell Pathway/ Category	Hedgehog,Pathways in cancer,Basal cell carcinoma,
Protein Expression	Fetal lung,Plasma,
Subcellular Localization	extracellular region,proteinaceous extracellular matrix,extracellular space,endoplasmic reticulum lumen,cytosol,plasma membrane,cell surface,membrane raft,

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Protein 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 N-terminus. N-product is active as a multimer.,tissue specificity:Expressed in fetal intestine, liver, lung, and kidney. Not expressed in adult tissues.,
Usage	For Research Use Only! Not for diagnostic or therapeutic procedures.