

# Rabbit Anti-IHH antibody

SL6624R

Product Name:	ІНН
Chinese Name:	HHG2抗体 MARKAN AND AND AND AND AND AND AND AND AND A
Alias:	HHG2; Indian Hedgehog; CENPB_HUMAN; Major centromere autoantigen B; Centromere protein B; CENP-B.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Horse,Rabbit,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500 (Paraffin sections
	need antigen repair)
	not vet tested in other applications.
	optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	45kDa
<b>Cellular localization:</b>	The cell membraneExtracellular matrixSecretory protein
Form:	Lyophilized or Liquid
<b>Concentration:</b>	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human IHH:161-260/411
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:	Ihh is an intercellular signalling peptide essential for a variety of patterning events during development. It binds to the patched (PTC) receptor, which functions in association with smoothened (SMO), to activate the transcription of target genes. Ihh induces the expression of parathyroid hormone-related protein (PTHRP) and is implicated in endochondral ossification, possibly regulating the balance between growth and ossification of the developing bones.

### Function:

Intercellular signal essential for a variety of patterning events during development. Binds to the patched (PTC) receptor, which functions in association with smoothened (SMO), to activate the transcription of target genes. Implicated in endochondral ossification: may regulate the balance between growth and ossification of the developing bones. Induces the expression of parathyroid hormone-related protein (PTHRP) (By similarity).

#### Subcellular Location:

Indian hedgehog protein N-product: Cell membrane; Lipid-anchor; Extracellular side (By similarity). Note=The N-terminal peptide remains associated with the cell surface (By similarity).

Indian hedgehog protein C-product: Secreted, extracellular space (By similarity). Note=The C-terminal peptide diffuses from the cell (By similarity).

#### **Tissue Specificity:**

Expressed in embryonic lung, and in adult kidney and liver.

#### **Post-translational modifications:**

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 (By similarity).

Cholesterylation is required for N-product targeting to lipid rafts and multimerization (By similarity).

Palmitoylated. N-palmitoylation is required for N-product multimerization and full activity.

## DISEASE:

Defects in IHH are the cause of brachydactyly type A1 (BDA1) [MIM:112500]. BDA1 is an autosomal dominant disorder characterized by middle phalanges of all the digits rudimentary or fused with the terminal phalanges. The proximal phalanges of the thumbs and big toes are short.

Defects in IHH are a cause of acrocapitofemoral dysplasia (ACFD) [MIM:607778]. ACFD is a disorder characterized by short stature of variable severity with postnatal onset. The most constant radiographic abnormalities are observed in the tubular bones of the hands and in the proximal part of the femur. Cone-shaped epiphyses or a similar epiphyseal configuration with premature epimetaphyseal fusion result in shortening of the skeletal components involved. Cone-shaped epiphyses were also present to a variable extent at the shoulders, knees, and ankles.

Similarity:

Belongs to the hedgehog family.

SWISS:

	P07199
	Gene ID: 3549
	Database links:
	Entrez Gene: 3549Human
	Entrez Gene: 16147 Mouse
	Entrez Gene: 84399Rat
	<u>Omim: 600726</u> Human
	SwissProt: Q14623Human
	SwissProt: P97812Mouse
	Unigene: 654504Human
	Unigene: 439736Mouse
	Unigene: 8711Rat
	<b>Important Note:</b> This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	

Paraformaldehyde-fixed, paraffin embedded (Rat liver); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (IHH) Polyclonal Antibody, Unconjugated (SL6624R) at 1:500 overnight at 4°C, followed by a conjugated secondary (sp-0023) for 20 minutes and DAB staining.

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