10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

Noggin Rabbit pAb

Catalog # AP98063

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q13253.1
Other Accession	Q13253.1
Reactivity	Rat, Human
Predicted	Pig, Mouse, Rabbit, Chicken, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	23 KDa
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Noggin
Epitope Specificity	28-130/232
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the noggin family.
SUBUNIT	Homodimer.
DISEASE	Defects in NOG are a cause of symphalangism proximal syndrome (SYM1) [MIM:185800]. SYM1 is characterized by the hereditary absence of the proximal interphalangeal (PIP) joints (Cushing symphalangism). Severity of PIP joint involvement diminishes towards the radial side. Distal interphalangeal joints are less frequently involved and metacarpophalangeal joints are rarely affected whereas carpal bone malformation and fusion are common. In the lower extremities, tarsal bone coalition is common. Conducive hearing loss is seen and is due to fusion of the stapes to the petrous part of the temporal bone.Defects in NOG are the cause of multiple synostoses syndrome type 1 (SYNS1) [MIM:186500]; also known as synostoses, multiple, with brachydactyly/symphalangism-brachydactyly syndrome. SYNS1 is characterized by tubular-shaped (hemicylindrical) nose with lack of alar flare, otosclerotic deafness, and multiple progressive joint fusions commencing in the hand. The joint fusions are progressive, commencing in the fifth proximal interphalangeal joint in early childhood (or at birth in some individuals) and progressing in an ulnar-to-radial and proximal-to-distal direction. With increasing age, ankylosis of other joints, including the cervical vertebrae, hips, and humeroradial joints, develop.Defects in NOG are the cause of tarsal-carpal coalition syndrome (TCC) [MIM:186570]. TCC is an autosomal dominant disorder characterized by fusion of the carpals, tarsals and phalanges, short first metacarpals causing brachydactyly, and humeroradial fusion. TCC is allelic to SYM1, and different mutations in NOG can result in either TCC or SYM1 in different families.Defects in NOG are a cause of stapes ankylosis with broad thumb and toes (SABTS) [MIM:184460]; also known as Teunissen-Cremers syndrome. SABTS is a congenital autosomal dominant disorder that includes hyperopia, a hemicylindrical nose, broad thumbs, great toes, and other minor skeletal anomalies but lacked carpal and tarsal fusion and symphalangism.Defects in NOG are the cause of brachydactyly type B2 (BDB2) [MIM:611377]. BDB2 is a

	subtype of brachydactyly characterized by hypoplasia/aplasia of distal phalanges in combination with distal symphalangism, fusion of carpal/tarsal bones, and partial cutaneous syndactyly.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Noggin is a secreted protein involved at multiple stages of vertebrate embryonic development including neural induction and is known to exert its effects by inhibiting the bone morphogenetic protein (BMP)-signaling pathway. It binds several BMPs with very high (picomolar) affinities, with a marked preference for BMP2 and BMP4 over BMP7. By binding tightly to BMPs, Noggin prevents BMPs from binding their receptors. Noggin binds the bone morphogenetic proteins (BMP) such as BMP-4 and BMP-7, and inhibits BMP signaling by blocking the molecular interfaces of the binding epitopes for both type I and type II receptors. Interaction of BMP and its antagonist Noggin governs various developmental and cellular processes, including embryonic dorsal-ventral axis, induction of neural tissue, formation of joints in the skeletal system and neurogenesis in the adult brain. Noggin plays a key role in neural induction by inhibiting BMP4, along with other TGF-β signaling inhibitors such as chordin and follistatin. Mouse knockout experiments have demonstrated that noggin also plays a crucial role in bone development, joint formation, and neural tube fusion.

Additional Information

Gene ID	9241
Other Names	SYM1; SYNS1; SYNS1A; noggin; synostoses (multiple) syndrome 1; symphalangism 1 (proximal)
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,Flow-Cyt=1ug/Test
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	Noggin Rabbit pAb
Function	Essential for cartilage morphogenesis and joint formation. Inhibitor of bone morphogenetic proteins (BMP) signaling which is required for growth and patterning of the neural tube and somite

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.