SunLong Biotech Co.,LTD
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**[ Primary Antibody ]**

# Rabbit Anti-Gli3 antibody

**Catalog Number: SL11565R**

**FOR RESEARCH USE ONLY**

**[ DATASHEET ]**

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| **Host:** | Rabbit |
| **Isotype:** | IgG |
| **Clonality:** | Polyclonal |
| **Target:** | Gli3 |
| **Immunogen:** | KLH conjugated synthetic peptide derived from human Gli3: 481-570/1580 |
| **Purification:** | affinity purified by Protein A |
| **Concentration:** | 1mg/ml |
| **Storage:** | 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. |
| **Applications:** | IHC-P=1:100-500<br>IHC-F=1:100-500<br>IF=1:100-500 （Paraffin sections need antigen repair）<br>not yet tested in other applications.<br>optimal dilutions/concentrations should be determined by the end user. |
| **Reactivity:** | Mouse,Rat<br>(predicted:Human,Chicken,Cow,Horse,Rabbit,Sheep) |
| **Predicted MW.:** | 170kDa |
| **Subcellular Location:** | The nucleus cytoplasmic |
| **Background:** | It has long been known that the overexpression of either Wnt-1 or the GLI proteins results in cancer; however, the molecular basis for this transformation was poorly understood. The Wnt-1 and GLI proteins have now been placed in a signaling cascade downstream of the mammalian homologs of the Drosophila hedgehog and patched proteins. The Drosophila segment polarity gene hedgehog (hh) encodes a secreted protein that appears to function in embryonic and imaginal disc patterning.<br>The ptc gene, also identified as a Drosophila segment polarity gene, encodes the transmembrane protein patched, the expression of which is precisely regulated during embryonic development. Hedgehog has been shown to enhance the expression of the Wnt family of proteins through a signaling cascade involving the GLI transcription factors, while patched functions as a |

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| | repressor opposing the effects of hedgehog. Mutations in the ptc gene, which result in unregulated hedgehog signaling, have been correlated with the most common type of cancer, basal cell carcinoma, which affects 750,000 individuals annually in the United States alone.<br>**Function:** Has a dual function as a transcriptional activator and a repressor of the sonic hedgehog (Shh) pathway, and plays a role in limb development. The full-length GLI3 form (GLI3FL) after phosphorylation and nuclear translocation, acts as an activator (GLI3A) while GLI3R, its C-terminally truncated form, acts as a repressor. A proper balance between the GLI3 activator and the repressor GLI3R, rather than the repressor gradient itself or the activator/repressor ratio gradient, specifies limb digit number and identity. In concert with TRPS1, plays a role in regulating the size of the zone of distal chondrocytes, in restricting the zone of PTHLH expression in distal cells and in activating chondrocyte proliferation. Binds to the minimal GLI-consensus sequence 5'-GGGTGGTC-3'.<br>**Subunit:** The full-length GLI3 form (GLI3FL) interacts with SUFU and this interaction regulates the formation of either repressor or activator forms of GLI3. Its association with SUFU is regulated by Hh signaling and dissociation of the SUFU-GLI3 interaction requires the presence of the ciliary motor KIF3A (By similarity). Interacts with KIF7. The activator form of GLI3 (GLI3A) but not the repressor form (GLI3R) can interact with TRPS1. The phosphorylated form interacts with BTRC. Interacts with ZIC1. Interacts with ZIC3 (via C2H2-type domains 3, 4 and 5); the interaction enhances its transcriptional activity.<br>**Subcellular Location:** Nucleus. Cytoplasm. Cell projection<br>**Tissue Specificity:** Is expressed in a wide variety of normal adult tissues, including lung, colon, spleen, placenta, testis, and myometrium.<br>**Post-translational modifications:** Phosphorylated on multiple sites by protein kinase A (PKA) and phosphorylation by PKA primes further phosphorylation by CK1 and GSK3. Phosphorylation is essential for its proteolytic processing. Transcriptional repressor GLI3R, a C-terminally truncated form, is generated from the full-length GLI3 protein (GLI3FL/GLI3-190) through proteolytic processing. This process requires PKA-primed phosphorylation of GLI3, ubiquitination of GLI3 and the presence of BTRC. GLI3FL is complexed with SUFU in the cytoplasm and is maintained in a neutral state.<br>Without the Hh signal, the SUFU-GLI3 complex is recruited to cilia, leading to the efficient processing of GLI3FL into GLI3R. GLI3R formation leads to its dissociation from SUFU, allowing it to translocate into the nucleus, and repress Hh target genes. When Hh signaling is initiated, SUFU dissociates from GLI3FL and this has two consequences. |

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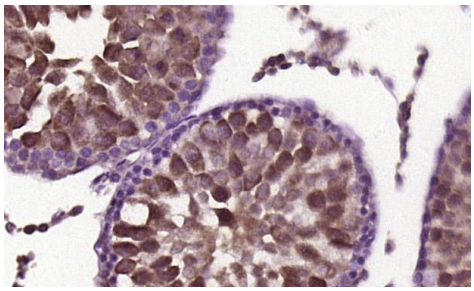
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| | First, GLI3R production is halted. Second, free GLI3FL translocates to the nucleus, where it is phosphorylated, destabilized, and converted to a transcriptional activator (GLI3A). Phosphorylated in vitro by ULK3.<br>DISEASE: Defects in GLI3 are the cause of Greig cephalo-poly-syndactyly syndrome (GCPS) [MIM:175700]. GCPS is an autosomal dominant disorder affecting limb and craniofacial development. It is characterized by pre- and postaxial polydactyly, syndactyly of fingers and toes, macrocephaly and hypertelorism.<br>Defects in GLI3 are a cause of Pallister-Hall syndrome (PHS) [MIM:146510]. PHS is characterized by a wide range of clinical manifestations. It mainly associates central or postaxial polydactyly, syndactyly, and hypothalamic hamartoma. Malformations are frequent in the viscera, e.g. anal atresia, bifid uvula, congenital heart malformations, pulmonary or renal dysplasia. It is an autosomal dominant disorder.<br>Defects in GLI3 are a cause of type A1/B postaxial polydactyly (PAPA1/PAPB) [MIM:174200, 603596]. PAPA in humans is an autosomal dominant trait characterized by an extra digit in the ulnar and/or fibular side of the upper and/or lower extremities. The extra digit is well formed and articulates with the fifth, or extra, metacarpal/metatarsal, and thus it is usually functional.<br>Defects in GLI3 are a cause of polydactyly preaxial type 4 (POP4) [MIM:174700]. Polydactyly preaxial type 4 (i.e., polydactyly on the radial/tibial side of the hand/foot) covers a heterogeneous group of entities. In preaxial polydactyly type IV, the thumb shows only the mildest degree of duplication, and syndactyly of various degrees affects fingers 3 and 4.<br>Defects in GLI3 are the cause of acrocallosal syndrome (ACS) [MIM:200990]; also abbreviated ACLS. ACS is characterized by postaxial polydactyly, hallux duplication, macrocephaly, and absence of the corpus callosum, usually with severe developmental delay.<br>**Similarity:** Belongs to the GLI C2H2-type zinc-finger protein family. Contains 5 C2H2-type zinc fingers.<br>**Database links:**<br>**Entrez Gene:** 420769 Chicken<br>**Entrez Gene:** 463369 Chimpanzee<br>**Entrez Gene:** 483244 Dog<br>**Entrez Gene:** 2737 Human<br>**Entrez Gene:** 14634 Mouse<br>**Entrez Gene:** 140588 Rat<br>Omim: 165240 Human<br>**SwissProt:** Q9IA31 Chicken |

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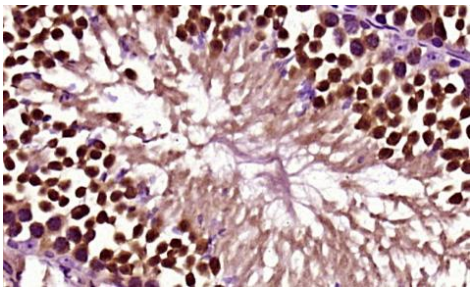
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| | **SwissProt:** Q5IS56 Chimpanzee<br>**SwissProt:** P10071 Human<br>**SwissProt:** Q61602 Mouse<br>**SwissProt:** Q91660 Xenopus laevis<br>**Unigene:** 21509 Human<br>**Unigene:** 5098 Mouse |

## [ VALIDATION IMAGES ]



Paraformaldehyde-fixed, paraffin embedded (rat testis tissue); 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 (Gli3) Polyclonal Antibody, Unconjugated (SL11565R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.



Paraformaldehyde-fixed, paraffin embedded (Mouse testis); 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 (Gli3) Polyclonal Antibody, Unconjugated (SL11565R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

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