




Anti-Gli3 antibody ab6050

★★★★★ [3 Abreviews](#) [22 References](#) [2 图像](#)

概述

产品名称	Anti-Gli3抗体
描述	兔多克隆抗体to Gli3
宿主	Rabbit
经测试应用	适用于: ICC/IF, IHC-Fr, WB, IHC-P
种属反应性	与反应: Mouse, Human 预测可用于: Rat, Chicken, Xenopus laevis 
免疫原	Synthetic peptide: NEDESPGQTYHRERRNA-C conjugated to KLH, corresponding to amino acids 41-57 of Human GLI3.  Run BLAST with  Run BLAST with
常规说明	<p>The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing.</p> <p>If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As</p>

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
存储溶液	Preservative: 0.01% Sodium azide Constituents: 0.42% Potassium phosphate, 0.87% Sodium chloride
纯度	Immunogen affinity purified
克隆	多克隆
同种型	IgG

应用

The Abpromise guarantee **Abpromise™**承诺保证使用ab6050于以下的经测试应用

“应用说明”部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
ICC/IF	★★★★★ (1)	Use a concentration of 1 µg/ml. See Abreview.
IHC-Fr	★★★★★ (1)	Use at an assay dependent concentration.
WB		Use at an assay dependent concentration. Predicted molecular weight: 170 kDa. PubMed: 23293081
IHC-P		Use a concentration of 0.5 - 5 µg/ml.

靶标

功能	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'.
组织特异性	Is expressed in a wide variety of normal adult tissues, including lung, colon, spleen, placenta, testis, and myometrium.
疾病相关	<p>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.</p> <p>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.</p> <p>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.</p> <p>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.</p> <p>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.</p>
序列相似性	<p>Belongs to the GLI C2H2-type zinc-finger protein family.</p> <p>Contains 5 C2H2-type zinc fingers.</p>

翻译后修饰

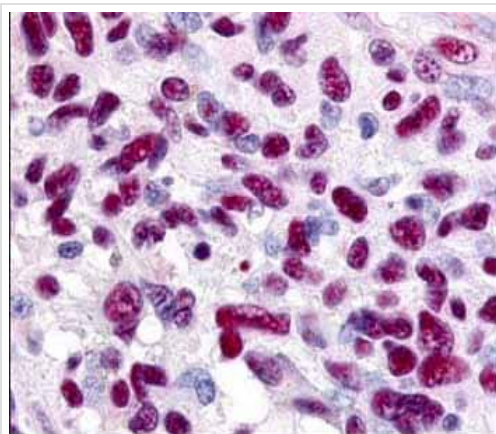
Phosphorylated on multiple sites by protein kinase A (PKA) and phosphorylation by PKA primes further phosphorylation by CK1 and GSK3. Phosphorylation is essential for its proteolytic processing.

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. 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. 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.

细胞定位

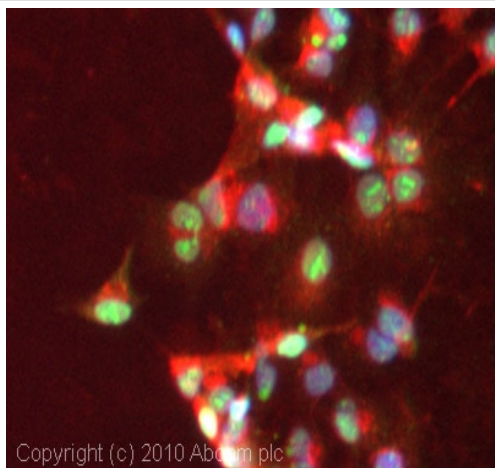
Nucleus. Cytoplasm. Cell projection > cilium. GLI3FL is localized predominantly in the cytoplasm while GLI3R resides mainly in the nucleus. Ciliary accumulation requires the presence of KIF7 and SMO. Translocation to the nucleus is promoted by interaction with ZIC1.

图片



ab6050 at 0.625 µg/ml staining human glioblastoma.

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Gli3 antibody (ab6050)



Immunocytochemistry/ Immunofluorescence - Anti-Gli3 antibody (ab6050)

ICC/IF image of ab6050 stained HepG2 cells. The cells were 4% formaldehyde fixed (10 min) and then incubated in 1%BSA / 10% normal goat serum / 0.3M glycine in 0.1% PBS-Tween for 1h to permeabilise the cells and block non-specific protein-protein interactions. The cells were then incubated with the antibody (ab6050, 1µg/ml) overnight at +4°C. The secondary antibody (green) was Alexa Fluor® 488 goat anti-rabbit IgG (H+L) used at a 1/1000 dilution for 1h. Alexa Fluor® 594 WGA was used to label plasma membranes (red) at a 1/200 dilution for 1h. DAPI was used to stain the cell nuclei (blue) at a concentration of 1.43µM.

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