

Product name:	GLI-3 Rabbit Polyclonal Antibody
Cat number:	ABN11465
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human GLI-3. AA range:11-60
Reactivity:	Human,Mouse,Rat
Applications:	IHC 1:100-1:300,ICC/IF 1:200-1:1000,ELISA 1:5000-1:20000
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

This gene encodes a protein which belongs to the C2H2-type zinc finger proteins subclass of the Gli family. They are characterized as DNA-binding transcription factors and are mediators of Sonic hedgehog (Shh) signaling. The protein encoded by this gene localizes in the cytoplasm and activates patched Drosophila homolog (PTCH) gene expression. It is also thought to play a role during embryogenesis. Mutations in this gene have been associated with several diseases, including Greig cephalopolysyndactyly syndrome, Pallister-Hall syndrome, preaxial polydactyly type IV, and postaxial polydactyly types A1 and B. [provided by RefSeq, Jul 2008],disease:Defects in GLI3 are a cause of Pallister-Hall syndrome (PHS) [MIM:146510]. Pallister-Hall syndrome 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.,disease: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.,disease:Defects in GLI3 are a cause of type IV preaxial polydactyly [MIM:174700]. Preaxial polydactyly (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.,disease: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.,disease:Defects in GLI3 are the cause of Greig cephalo-poly-syndactyly syndrome (GCPS) [MIM:175700]; an autosomal dominant disorder affecting limb and craniofacial development. GCPS is characterized by pre- and postaxial polydactyly, syndactyly of fingers and toes, macrocephaly and hypertelorism.,function:Plays a role in limb and brain development. Implicated in the transduction of SHH signal.,similarity:Belongs to the GLI C2H2-type zinc-finger protein family.,similarity:Contains 5 C2H2-type zinc fingers.,tissue specificity:Is expressed in a wide variety of normal adult tissues, including lung, colon, spleen, placenta, testis, and myometrium.,