



Rabbit Anti-FGFR1 antibody

SL20226R

Product Name:	FGFR1
Chinese Name:	碱性成纤维细胞生长因子受体1抗体
Alias:	bFGF R; BFGFR; C FGR; CD 331; CD331; CD331 antigen; CEK; FGFBR; FGFR 1; Fibroblast growth factor receptor 1; FLG; FLG protein; FLJ14326; FLT 2; FLT2; Fms like tyrosine kinase 2; Fms related tyrosine kinase 2; Fms related tyrosine kinase 2 Pfeiffer syndrome; H2; H3; H4; H5; HBGFR; Heparin binding growth factor receptor; Hydroxyaryl protein kinase; KAL 2; KAL2; MFR; N SAM; N sam tyrosine kinase; Protein tyrosine kinase; Tyrosylprotein kinase; Basic fibroblast growth factor receptor 1; FGFR1 HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,
Applications:	ELISA=1:500-1000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	88kDa
Cellular localization:	The nucleuscytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human FGFR1:31-130/822<Extracellular>
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:	Fibroblast growth factors (FGFs) produce mitogenic and angiogenic effects in target

cells by signaling through the cellular surface tyrosine kinase receptors. There are four members of the FGF receptor family: FGFR-1 (flg), FGFR-2 (bek, KGFR), FGFR-3 and FGFR-4. Each receptor contains an extracellular ligand binding domain, a transmembrane region and a cytoplasmic kinase domain (1). Following ligand binding and dimerization, the receptors are phosphorylated at specific tyrosine residues (2). Seven tyrosine residues in the cytoplasmic tail of FGFR-1 can be phosphorylated: Tyr463, Tyr583, Tyr585, Tyr653, Tyr654, Tyr730 and Tyr766. Tyrosine 653 and 654 are important for catalytic activity of the activated FGFR and are essential for signaling (3). The other phosphorylated tyrosine residues may provide docking sites for downstream signaling components such as Crk and PLCgamma.

Function:

Receptor for fibroblast growth factors FGF2 and FGF1. Receptor for FGF23 in the presence of KL (By similarity). Promotes mitogenesis in response to fibroblast growth factors. Activates PLCG1.

Subunit:

Homodimer. Interacts with KLB. Interacts with KL and FGF23. Interacts with SHB and GRB10. Interacts with PLCG1 (via SH2 domains). Interacts with KAL1; this interaction does not interfere with FGF2-binding to FGFR1, but prevents binding of heparin-bound FGF2. Interacts with SOX2 and SOX3.

Subcellular Location:

Membrane; Single-pass type I membrane protein.

Post-translational modifications:

Binding of FGF1 and heparin promotes autophosphorylation on tyrosine residues and activation of the receptor.

DISEASE:

Defects in FGFR1 are a cause of Pfeiffer syndrome (PS) [MIM:101600]; also known as acrocephalosyndactyly type V (ACS5). PS is characterized by craniosynostosis (premature fusion of the skull sutures) with deviation and enlargement of the thumbs and great toes, brachymesophalangy, with phalangeal ankylosis and a varying degree of soft tissue syndactyly.

Defects in FGFR1 are a cause of idiopathic hypogonadotropic hypogonadism (IHH) [MIM:146110]. IHH is defined as a deficiency of the pituitary secretion of follicle-stimulating hormone and luteinizing hormone, which results in the impairment of pubertal maturation and of reproductive function.

Defects in FGFR1 are the cause of Kallmann syndrome type 2 (KAL2) [MIM:147950]; also known as hypogonadotropic hypogonadism and anosmia. Anosmia or hyposmia is related to the absence or hypoplasia of the olfactory bulbs and tracts. Hypogonadism is due to deficiency in gonadotropin-releasing hormone and probably results from a failure of embryonic migration of gonadotropin-releasing hormone-synthesizing neurons. In some cases, midline cranial anomalies (cleft lip/palate and imperfect fusion) are present and anosmia may be absent or inconspicuous.

Defects in FGFR1 are the cause of osteoglophonic dysplasia (OGD) [MIM:166250]; also known as osteoglophonic dwarfism. OGD is characterized by craniosynostosis, prominent supraorbital ridge, and depressed nasal bridge, as well as by rhizomelic dwarfism and nonossifying bone lesions. Inheritance is autosomal dominant.

Defects in FGFR1 are the cause of trigonocephaly non-syndromic (TRICEPH) [MIM:190440]; also known as metopic craniosynostosis. The term trigonocephaly describes the typical keel-shaped deformation of the forehead resulting from premature fusion of the frontal suture. Trigonocephaly may occur also as a part of a syndrome.

Note=A chromosomal aberration involving FGFR1 may be a cause of stem cell leukemia lymphoma syndrome (SCLL). Translocation t(8;13)(p11;q12) with ZMYM2. SCLL usually presents as lymphoblastic lymphoma in association with a myeloproliferative disorder, often accompanied by pronounced peripheral eosinophilia and/or prominent eosinophilic infiltrates in the affected bone marrow.

[DISEASE] Note=A chromosomal aberration involving FGFR1 may be a cause of stem cell myeloproliferative disorder (MPD). Translocation t(6;8)(q27;p11) with FGFR1OP. Insertion ins(12;8)(p11;p11p22) with FGFR1OP2. MPD is characterized by myeloid hyperplasia, eosinophilia and T-cell or B-cell lymphoblastic lymphoma. In general it progresses to acute myeloid leukemia. The fusion proteins FGFR1OP2-FGFR1, FGFR1OP-FGFR1 or FGFR1-FGFR1OP may exhibit constitutive kinase activity and be responsible for the transforming activity.

Note=A chromosomal aberration involving FGFR1 may be a cause of stem cell myeloproliferative disorder (MPD). Translocation t(8;9)(p12;q33) with CEP110. MPD is characterized by myeloid hyperplasia, eosinophilia and T-cell or B-cell lymphoblastic lymphoma. In general it progresses to acute myeloid leukemia. The fusion protein CEP110-FGFR1 is found in the cytoplasm, exhibits constitutive kinase activity and may be responsible for the transforming activity.

Similarity:

Belongs to the protein kinase superfamily. Tyr protein kinase family. Fibroblast growth factor receptor subfamily.
Contains 3 Ig-like C2-type (immunoglobulin-like) domains.
Contains 1 protein kinase domain.

SWISS:

P11362

Gene ID:

2260

Database links:

[Entrez Gene: 2260](#) Human

[Entrez Gene: 14182](#) Mouse

[Entrez Gene: 79114](#) Rat

[Omim: 136350](#) Human

[SwissProt: P11362](#) Human

[SwissProt: P16092](#) Mouse

[SwissProt: Q04589](#) Rat

[Unigene: 264887](#) Human

[Unigene: 265716](#) Mouse

[Unigene: 207203](#) Rat

[Unigene: 9797](#) Rat

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

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