

FGF Receptor 1 Recombinant Rabbit mAb

Catalog # AP100319

Product Information

Application	WB, IHC-P, IHC-F, IF, IP, E
Primary Accession	P11362
Other Accession	P11362
Reactivity	Rat, Human, Mouse
Host	Rabbit
Clonality	Recombinant
Calculated MW	91868
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 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.
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.
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.
Background Descriptions	The protein encoded by this gene is a member of the fibroblast growth factor receptor (FGFR) family, where amino acid sequence is highly conserved between members and throughout evolution. FGFR family members differ from one another in their ligand affinities and tissue distribution. A full-length representative protein consists of an extracellular region, composed of three immunoglobulin-like domains, a single hydrophobic membrane-spanning segment and a cytoplasmic tyrosine kinase domain. The extracellular portion of the protein interacts with fibroblast growth factors, setting in motion a cascade of downstream signals, ultimately influencing mitogenesis and differentiation. This particular family member binds both acidic and basic fibroblast growth factors and is involved in limb induction. Mutations in this gene have been associated with Pfeiffer syndrome, Jackson-Weiss syndrome, Antley-Bixler syndrome, osteoglophonic dysplasia, and autosomal dominant Kallmann syndrome 2. Chromosomal aberrations involving this gene are associated with stem cell myeloproliferative disorder and stem cell leukemia lymphoma syndrome. Alternatively spliced variants which encode different protein isoforms have been described; however, not all variants have been

Additional Information

Gene ID	2260
Other Names	BFGFR; CD331; CEK; ECCL; FGFBR; FGFR-1; FLG; FLT-2; FLT2; HBGFR; HH2; HRTFDS; KAL2; N-SAM; OGD; bFGF-R-1; Eask; FGFR-I; Fr1; Hspy; MFR; c-fgr; bFGF-R; FGFR1_HUMAN; FGFR1; Basic fibroblast growth factor receptor 1 (BFGFR bFGF-R-1); Fms-like tyrosine kinase 2 (FLT-2); Proto-oncogene c-Fgr; 2.7.10.1; FGFR1_MOUSE; Basic fibroblast growth factor receptor 1; FGFR1_RAT; fibroblast growth factor receptor 1; fms-related tyrosine kinase 2; Pfeiffer syndrome; Fms-like tyrosine kinase 2; CD antigen CD331
Dilution	IHC-P=1:50-200,IHC-F=1:50-200,WB=1:500-2000,IF=1:50-200,ELISA=1:5000-10 000,IP=1:50-200
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	FGFR1
Synonyms	BFGFR, CEK, FGFBR, FLG, FLT2, HBGFR
Function	Tyrosine-protein kinase that acts as a cell-surface receptor for fibroblast growth factors and plays an essential role in the regulation of embryonic development, cell proliferation, differentiation and migration. Required for normal mesoderm patterning and correct axial organization during embryonic development, normal skeletogenesis and normal development of the gonadotropin-releasing hormone (GnRH) neuronal system. Phosphorylates PLCG1, FRS2, GAB1 and SHB. Ligand binding leads to the activation of several signaling cascades. Activation of PLCG1 leads to the production of the cellular signaling molecules diacylglycerol and inositol 1,4,5-trisphosphate. Phosphorylation of FRS2 triggers recruitment of GRB2, GAB1, PIK3R1 and SOS1, and mediates activation of RAS, MAPK1/ERK2, MAPK3/ERK1 and the MAP kinase signaling pathway, as well as of the AKT1 signaling pathway. Promotes phosphorylation of SHC1, STAT1 and PTPN11/SHP2. In the nucleus, enhances RPS6KA1 and CREB1 activity and contributes to the regulation of transcription. FGFR1 signaling is down-regulated by IL17RD/SEF, and by FGFR1 ubiquitination, internalization and degradation.
Cellular Location	Cell membrane; Single-pass type I membrane protein. Nucleus. Cytoplasm, cytosol. Cytoplasmic vesicle. Note=After ligand binding, both receptor and ligand are rapidly internalized. Can translocate to the nucleus after internalization, or by translocation from the endoplasmic reticulum or Golgi apparatus to the cytosol, and from there to the nucleus
Tissue Location	Detected in astrocytoma, neuroblastoma and adrenal cortex cell lines. Some isoforms are detected in foreskin fibroblast cell lines, however isoform 17, isoform 18 and isoform 19 are not detected in these cells.

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