

Rabbit Anti-EWSR1 antibody

SL10552R

Product Name:	EWSR1
Chinese Name:	尤文氏肉瘤相关EWS蛋白抗体
Alias:	bK984G1.4; bK984G1.4 Ewing sarcoma breakpoint region 1 protein; Ewing sarcoma breakpoint region 1; Ewing sarcoma breakpoint region 1 protein; Ewings sarcoma EWS Fli1 type 1 oncogene; EWS; EWS oncogene; EWS_HUMAN; EWSR 1; Ewsr1; EWSR1 protein; RNA binding protein EWS; RNA-binding protein EWS.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human, Mouse, Rat, Dog, Pig, Cow, Rabbit,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	68kDa
Cellular localization:	The nucleuscytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	lmg/ml
immunogen:	KLH conjugated synthetic peptide derived from human EWSR1:351-450/656
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:	<u>PubMed</u>
Product Detail:	EWS is a nuclear RNA-binding protein. As a result of chromosome translocation, the EWS gene is fused to a variety of transcription factors, including ATF-1 in human neoplasias. In the Ewing family of tumors, the N-terminal domain of EWS is fused to the DNA-binding domain of various ETS transcription factors, including Fli-1, Erg,

ETV1, E1AF and FEV. The EWS/Fli-1 chimeric protein acts as a more potent transcriptional activator than Fli-1 and can promote cell transformation. Two functional regions have been identified in EWS; an amino-terminal region (domain A), that has little transactivation activity but transforms efficiently when fused to Fli-1, and a distal region (domain B), which shows transactivation activity but transforms less efficiently when fused to Fli-1.

Function:

Might normally function as a repressor. EWS-fusion-proteins (EFPS) may play a role in the tumorigenic process. They may disturb gene expression by mimicking, or interfering with the normal function of CTD-POLII within the transcription initiation complex. They may also contribute to an aberrant activation of the fusion protein target genes.

Subunit:

Binds POLR2C, SF1, calmodulin and RNA. Interacts with PTK2B/FAK2 and TDRD3. Binds calmodulin in the presence, but not in the absence, of calcium ion.

Subcellular Location:

Nucleus. Cytoplasm. Cell membrane. Relocates from cytoplasm to ribosomes upon PTK2B/FAK2 activation.

Tissue Specificity:

Ubiquitous.

Post-translational modifications:

Phosphorylated; calmodulin-binding inhibits phosphorylation of Ser-266. Highly methylated on arginine residues. Methylation is mediated by PRMT1 and, at lower level by PRMT8.

DISEASE:

Defects in EWSR1 are a cause of Ewing sarcoma (ES) [MIM:612219]. A highly malignant, metastatic, primitive small round cell tumor of bone and soft tissue that affects children and adolescents. It belongs to the Ewing sarcoma family of tumors, a group of morphologically heterogeneous neoplasms that share the same cytogenetic features. They are considered neural tumors derived from cells of the neural crest. Ewing sarcoma represents the less differentiated form of the tumors.

Note=Chromosomal aberrations involving EWSR1 are found in patients with Ewing sarcoma. Translocation t(11;22)(q24;q12) with FLI1; translocation t(7;22)(p22;q12) with ETV1; translocation t(21;22)(q22;q12) with ERG; translocation t(9;22)(q22-31;q11-12) with NR4A3. Translocation t(2;21;22)(q23;q22;q12) that forms a EWSR1-FEV fusion protein with potential oncogenic activity.

Note=A chromosomal aberration involving EWSR1 is associated with desmoplastic small round cell tumor (DSRCT). Translocation t(11;22)(p13;q12) with WT1. Note=A chromosomal aberration involving EWSR1 is associated with malignant melanoma of soft parts (MMSP). Translocation t(12;22)(q13;q12) with ATF-1.

Malignant melanoma of soft parts, also known as soft tissue clear cell sarcoma, is a rare tumor developing in tendons and aponeuroses.

Note=A chromosomal aberration involving EWSR1 is associated with small round cell sarcoma. Translocation t(11;22)(p36.1;q12) with PATZ1.

Defects in EWSR1 may be a cause of angiomatoid fibrous histiocytoma (AFH) [MIM:612160]. A distinct variant of malignant fibrous histiocytoma that typically occurs in children and adolescents and is manifest by nodular subcutaneous growth. Characteristic microscopic features include lobulated sheets of histiocyte-like cells intimately associated with areas of hemorrhage and cystic pseudovascular spaces, as well as a striking cuffing of inflammatory cells, mimicking a lymph node metastasis. Note=Chromosomal aberrations involving EWSR1 are found in patients with angiomatoid fibrous histiocytoma. Translocation t(12;22)(q13;q12) with ATF1 generates a chimeric EWSR1/ATF1 protein. Translocation t(2;22)(q33;q12) with CREB1 generates a EWSR1/CREB1 fusion gene that is most common genetic abnormality in this tumor type.

Note=EFPS arise due to chromosomal translocations in which EWSR1 is fused to a variety of cellular transcription factors. EFPS are very potent transcriptional activators dependent on the EAD and a C-terminal DNA-binding domain contributed by the fusion partner. The spectrum of malignancies associated with EFPS are thought to arise via EFP-induced transcriptional deregulation, with the tumor phenotype specified by the EWSR1 fusion partner and cell type. Transcriptional repression of the transforming growth factor beta type II receptor (TGF beta RII) is an important target of the EWS-FLI1, EWS-ERG, or EWS-ETV1 oncogene.

Similarity:

Belongs to the RRM TET family.

Contains 1 IQ domain.

Contains 1 RanBP2-type zinc finger.

Contains 1 RRM (RNA recognition motif) domain.

SWISS:

O01844

Gene ID:

2130

Database links:

Entrez Gene: 2130Human

Entrez Gene: 14030Mouse

Entrez Gene: 289752Rat

Omim: 133450Human

SwissProt: O01844Human

SwissProt: Q61545Mouse

Unigene: 374477Human

Unigene: 142822Mouse

Unigene: 52785Rat

Important Note:

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