

Estrogen Related Receptor beta Rabbit pAb

Catalog # AP98294

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	O95718
Other Accession	O95718
Host	Rabbit
Clonality	Polyclonal
Calculated MW	48054
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Estrogen Related Receptor beta
Epitope Specificity	131-230/508
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 nuclear hormone receptor family. NR3 subfamily. Contains 1 nuclear receptor DNA-binding domain.
SUBUNIT	Monomer.
DISEASE	Defects in ESRRB are the cause of deafness autosomal recessive type 35 (DFNB35) [MIM:608565]. DFNB35 is a form of sensorineural hearing loss. Sensorineural deafness results from damage to the neural receptors of the inner ear, the nerve pathways to the brain, or the area of the brain that receives sound information.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Estrogen related receptor beta, a NR3 Steroid Receptor, has been shown to affect early placental development. Also it can act as a repressor of transcriptional activity mediated by the glucocorticoid receptor (GR). It binds as a monomer to the extended half-site TNAAGTGCA and as a homodimer to the estrogen response element (ERE), palindromic thyroid hormone response element (TRE(pal)), and SF-1 response element, but not to the glucocorticoid response element (GRE). Estrogen related receptor beta mutations lead to abnormal development of the chorion and defective diploid trophoblast proliferation, and are lethal during midgestation. Expression has been documented in mice in the extraembryonic ectoderm that gives rise to the chorion. ESTs have been isolated from normal human eye, lung, and testis libraries. The ligand for the receptor is PPARgamma coactivator 1 (PGC-1) beta.

Additional Information

Gene ID	2103
Other Names	DFNB35; ERR beta-2; ERR2; ERRb; ERRbeta2; ESRL2; NR3B2; Estrrb; ERR2_HUMAN; ESRRB; Estrogen receptor-like 2; Estrogen-related receptor

beta (ERR-beta); Nuclear receptor subfamily 3 group B member 2; ERRB2; ERR2_MOUSE; Err-2; ERR2_RAT; estrogen related receptor beta; deafness, autosomal recessive 35

Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ELISA=1:5000-10000
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	ESRRB (HGNC:3473)
Synonyms	ERRB2, ESRL2, NR3B2
Function	[Isoform 3]: Transcription factor that binds a canonical ESRRB recognition (ERRE) sequence 5'TCAAGGTCA-3' localized on promoter and enhancer of targets genes regulating their expression or their transcription activity (PubMed: 17920186 , PubMed: 19755138). Plays a role, in a LIF-independent manner, in maintainance of self-renewal and pluripotency of embryonic and trophoblast stem cells through different signaling pathways including FGF signaling pathway and Wnt signaling pathways. Involved in morula development (2-16 cells embryos) by acting as a regulator at the 8-cell stage (By similarity). Upon FGF signaling pathway activation, interacts with KDM1A by directly binding to enhancer site of ELF5 and EOMES and activating their transcription leading to self-renewal of trophoblast stem cells. Also regulates expression of multiple rod-specific genes and is required for survival of this cell type (By similarity). Plays a role as transcription factor activator of GATA6, NR0B1, POU5F1 and PERM1 (PubMed: 23836911). Plays a role as transcription factor repressor of NFE2L2 transcriptional activity and ESR1 transcriptional activity (PubMed: 17920186 , PubMed: 19755138). During mitosis remains bound to a subset of interphase target genes, including pluripotency regulators, through the canonical ESRRB recognition (ERRE) sequence, leading to their transcriptional activation in early G1 phase. Can coassemble on structured DNA elements with other transcription factors like SOX2, POU5F1, KDM1A and NCOA3 to trigger ESRRB-dependent gene activation. This mechanism, in the case of SOX2 corecruitment prevents the embryonic stem cells (ESCs) to epiblast stem cells (EpiSC) transition through positive regulation of NR0B1 that inhibits the EpiSC transcriptional program. Also plays a role inner ear development by controlling expression of ion channels and transporters and in early placentation (By similarity).
Cellular Location	Nucleus. Cytoplasm {ECO:0000250 UniProtKB:Q61539}. Chromosome {ECO:0000250 UniProtKB:Q61539}

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