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Rabbit Anti-AIRE antibody

SL12453R

Product Name:	AIRE
Chinese Name:	免疫调节蛋白AIRE抗体
Alias:	AIRE-1; AIRE; AIRE_HUMAN; AIRE1; APECED; APECED protein; APS1; APSI; Autoimmune polyendocrinopathy candidiasis ectodermal dystrophy protein; Autoimmune regulator; Autoimmune regulator protein; PGA1.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Horse,
Applications:	ELISA=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:	58kDa
Cellular localization:	The nucleuscytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human PGA1/AIRE:51-150/545
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:	The autoimmune regulator gene, which is defective in the hereditary auto-immune disease APECED, encodes the transcriptional activator AIRE. AIRE is expressed in the medullary epithelial cells and monocyte-dendritic cells of the thymus, with lower expression in the spleen, fetal liver and lymph nodes. In adult tissue, AIRE expression in the thymus is confined to the medulla and the cortico-medullary junction, where it is

modulated by thymocytes undergoing negative selection. At the cellular level, AIRE is located in microtubular structures of the cyto-skeleton and in discrete nuclear dots resembling ND10 nuclear bodies. AIRE is induced by developing early thymocytes and is associated with the correct establishment of a regular thymic environment. AIRE regulates thymic architecture via transcriptional control of downstream target genes. AIRE mutations in APECED patients may affect thymic T cell selection and the formation of self-tolerance.

Function:
Transcriptional regulator that binds to DNA as a dimer or as a tetramer, but not as a monomer. Binds to G-doublets in an A/T-rich environment; the preferred motif is a tandem repeat of 5'-. ATTGGTTA-3' combined with a 5'-TTATTA-3' box. Binds to nucleosomes (By similarity). Binds to chromatin and interacts selectively with histone H3 that is not methylated at 'Lys-4', not phosphorylated at 'Thr-3' and not methylated at 'Arg-2'. Functions as a sensor of histone H3 modifications that are important for the epigenetic regulation of gene expression. Functions as a transcriptional activator and promotes the expression of otherwise tissue-specific self-antigens in the thymus, which is important for self tolerance and the avoidance of autoimmune reactions.

Subcellular Location:
Nucleus. Cytoplasm. Associated with tubular structures and in discrete nuclear dots resembling ND10 nuclear bodies. May shuttle between nucleus and cytoplasm.Target information above from: UniProt accession O43918 The UniProt Consortium The Universal Protein Resource (UniProt) in 2010 Nucleic Acids Res. 38:D142-D148 (2010) . Information by UniProt

Tissue Specificity:
Widely expressed. Expressed at higher level in thymus (medullary epithelial cells and monocyte-dendritic cells), pancreas, adrenal cortex and testis. Expressed at lower level in the spleen, fetal liver and lymph nodes. Isoform 2 and isoform 3 seem to be less frequently expressed than isoform 1, if at all.

Post-translational modifications:
Phosphorylated. Phosphorylation could trigger oligomerization.

DISEASE:
Defects in AIRE are a cause of autoimmune poly-endocrinopathy candidiasis ectodermal dystrophy (APECED) [MIM:240300]; also known as autoimmune polyglandular syndrome type I (APS-1). APECED is an autosomal recessive disease characterized by: (1) autoimmune polyendocrinopathies: hypoparathyroidism, adrenocortical failure, IDDM, gonadal failure, hypothyroidism, pernicious anemia, and hepatitis; (2) chronic mucocutaneous candidiasis; (3) ectodermal dystrophies: vitiligo, alopecia, keratopathy, dystrophy of dental enamel, nails and tympanic membranes. In addition, a high proportion of patients develop squamous cell carcinoma of the oral mucosa. The disease is reported worldwide but is exceptionally prevalent among the Finnish population (incidence 1:25000) and the Iranian jews (incidence 1:9000).

Note=Most of the mutations alter the nucleus-cytoplasm distribution of AIRE and disturb its association with nuclear dots and cytoplasmic filaments. Most of the mutations also decrease transactivation of the protein. The HSR domain is responsible for the homomultimerization activity of AIRE. All the missense mutations of the HSR and the SAND domains decrease this activity, but those in other domains do not. The AIRE protein is present in soluble high-molecular-weight complexes. Mutations in the HSR domain and deletion of PHD zinc fingers disturb the formation of these complexes.

Similarity:
Contains 1 HSR domain.
Contains 2 PHD-type zinc fingers.
Contains 1 SAND domain.

SWISS:
O43918

Gene ID:
326

Database links:

Entrez Gene: 326Human

Entrez Gene: 11634Mouse

Entrez Gene: 294328Rat

Entrez Gene: 487796Dog

Omim: 607358Human

SwissProt: O43918Human

SwissProt: Q9Z0E3Mouse

Unigene: 129829Human

Unigene: 35300Mouse

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