

Catalog # AF1041a

Goat Anti-AIRE (isoform 1) Antibody Peptide-affinity purified goat antibody

### Specification

## Goat Anti-AIRE (isoform 1) Antibody - Product Information

Application Primary Accession Other Accession Reactivity Predicted Host Clonality Concentration Isotype Calculated MW WB, IHC <u>O43918</u> <u>NP\_000374</u>, <u>326</u> Human, Pig Rat, Dog Goat Polyclonal 100ug/200ul IgG 57727

## Goat Anti-AIRE (isoform 1) Antibody - Additional Information

Gene ID 326

**Other Names** 

Autoimmune regulator, Autoimmune polyendocrinopathy candidiasis ectodermal dystrophy protein, APECED protein, AIRE, APECED

#### Format

0.5 mg lgG/ml in Tris saline (20mM Tris pH7.3, 150mM NaCl), 0.02% sodium azide, with 0.5% bovine serum albumin

Storage

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

**Precautions** Goat Anti-AIRE (isoform 1) Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

### Goat Anti-AIRE (isoform 1) Antibody - Protein Information

Name AIRE

Synonyms APECED

Function

Transcription factor playing an essential role to promote self-tolerance in the thymus by regulating the expression of a wide array of self-antigens that have the commonality of being tissue-restricted in their expression pattern in the periphery, called tissue restricted antigens (TRA)



(PubMed:<a href="http://www.uniprot.org/citations/26084028" target=" blank">26084028</a>). 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. Mainly expressed by medullary thymic epithelial cells (mTECs), induces the expression of thousands of tissue-restricted proteins, which are presented on major histocompatibility complex class I (MHC-I) and MHC-II molecules to developing T-cells percolating through the thymic medulla (PubMed: <a href="http://www.uniprot.org/citations/26084028" target="\_blank">26084028</a>). Also induces self- tolerance through other mechanisms such as the regulation of the mTEC differentiation program. Controls the medullary accumulation of thymic dendritic cells and the development of regulatory T-cell through the regulation of XCL1 expression. Regulates the production of CCR4 and CCR7 ligands in medullary thymic epithelial cells and alters the coordinated maturation and migration of thymocytes. In thimic B-cells, allows the presentation of licensing-dependent endogenous self-anitgen for negative selection. In secondary lymphoid organs, induces functional inactivation of CD4(+) T-cells. Expressed by a distinct bone marrow-derived population, induces self-tolerance through a mechanism that does not require regulatory T-cells and is resitant to innate inflammatory stimuli (By similarity).

#### **Cellular Location**

Nucleus. Cytoplasm. Note=Predominantly nuclear but also cytoplasmic (PubMed:11274163, PubMed:14974083). Found in nuclear body- like structures (dots) and in a filamentous vimentin-like pattern (PubMed:11274163, PubMed:14974083, PubMed:26084028). Associated with tubular structures (PubMed:11274163, PubMed:14974083)

### Tissue Location

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. In secondary lymphoid organs, expressed in a discrete population of bone marrow-derived toleregenic antigen presenting cells (APCs) called extrathymic AIRE expressing cells (eTAC)(at protein level) (PubMed:23993652). Isoform 2 and isoform 3 seem to be less frequently expressed than isoform 1, if at all

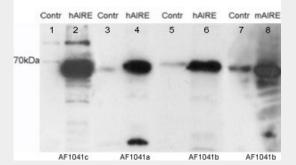
### Goat Anti-AIRE (isoform 1) Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

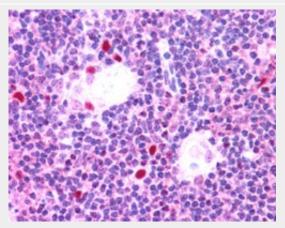
- <u>Western Blot</u>
- Blocking Peptides
- <u>Dot Blot</u>
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- <u>Cell Culture</u>

Goat Anti-AIRE (isoform 1) Antibody - Images





AF1041a staining (0.05  $\mu$ g/ml) of HEK293 cell lysates. Untransfected (Lane 3) and transfected with Human AIRE (lane 4). Data kindly provided by Prof. Pärt Peterson, University of Tartu, Estonia.



AF1041a (2.5  $\mu$ g/ml) staining of paraffin embedded Human Thymus. Steamed antigen retrieval with citrate buffer pH 6, AP-staining.

# Goat Anti-AIRE (isoform 1) Antibody - Background

This gene encodes a transcriptional regulator that forms nuclear bodies and interacts with the transcriptional coactivator CBP. At least three splice variant mRNAs products have been described including one which results in a premature stop codon and a transcript predicted to be a candidate for nuclear-mediated decay (NMD). Defects in this gene cause the rare autosomal-recessive systemic autoimmune disease termed autoimmune polyendocrinopathy-candidiasis-ectodermal dystrophy (APECED).

# Goat Anti-AIRE (isoform 1) Antibody - References

AIRE GENE MUTATIONS AND AUTOANTIBODIES TO INTERFERON OMEGA IN PATIENTS WITH CHRONIC HYPOPARATHYROIDISM WITHOUT APECED. Cervato S, et al. Clin Endocrinol (Oxf), 2010 Aug 13. PMID 20718774.

Physiogenomic analysis of statin-treated patients: domain-specific counter effects within the ACACB gene on low-density lipoprotein cholesterol? Rua
O G, et al. Pharmacogenomics, 2010 Jul. PMID 20602615.

The role of AIRE polymorphisms in melanoma. Conteduca G, et al. Clin Immunol, 2010 Jul. PMID 20363194.

Aire regulates the expression of differentiation-associated genes and self-renewal of embryonic stem cells. Gu B, et al. Biochem Biophys Res Commun, 2010 Apr 2. PMID 20226168.

Increased apoptosis after autoimmune regulator expression in epithelial cells revealed by a combined quantitative proteomics approach. Colom N, et al. J Proteome Res, 2010 May 7. PMID 20218732.