SunLong Biotech Co.,LTD
Tel: 0086-571- 56623320 Fax:0086-571- 56623318
E-mail:sales@sunlongbiotech.com
www.sunlongbiotech.com

Rabbit Anti-NR0B1 antibody

SL10434R

Product Name:	NR0B1
Chinese Name:	肾上腺发育不全相关蛋白抗体
Alias:	NR0B1 / Dax1; AHC; AHCH; AHX; DAX 1; DAX-1; DAX1; Dosage sensitive sex reversal; DSS; DSS AHC critical region on the X chromosome protein 1; DSS-AHC critical region on the X chromosome protein 1; GTD; HHG; Nr0b1; NR0B1_HUMAN; NROB1; Nuclear hormone receptor; Nuclear receptor 0B1; Nuclear receptor DAX 1; Nuclear receptor DAX-1; Nuclear receptor DAX1; Nuclear receptor subfamily 0 group B member 1; SRXY2.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Pig,Cow,Horse,Rabbit,Sheep,
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:	52kDa
Cellular localization:	The nucleuscytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human NR0B1:331-430/470
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:	Adrenal hypoplasia congentia (AHC) is an X-linked disorder characterized by primary adrenal insufficiency. The disorder, which is lethal if untreated, results in adrenal

	insufficiency early in infancy and is characterized by low serum concentration of glucocorticoids, mineralcorticoids and androgens and failure to respond to ACTH. AHC has been mapped to chromosome Xp21 at the same or close to an X-linked locus involved in sex determination, DSS (for dosage-sensitive sex reversal). The gene corresponding to DSS and AHC (designated DAX-1 for DSS-AHC critical region on the X chromosome, gene 1) has been cloned and shown to be deleted in AHC deletion patients and mutated in AHC non-deletion patients. The carboxy terminal 250 amino acids of the DAX-1-encoded protein, DAX-1, exhibits approximately 50% continuous similarity to the ligand-binding domain of the members of the nuclear hormone receptor superfamily while the amino terminal domain contains a putative DNA-binding motif. DAX-1 binds to retinoic acid responsive elements and down regulates retinoic acid receptor-mediated transcriptional activation. **Function:** Orphan nuclear receptor. Component of a cascade required for the development of the hypothalamic-pituitary-adrenal-gonadal axis. Acts as a coregulatory protein that inhibits the transcriptional activity of other nuclear receptors through heterodimeric interactions. May also have a role in the development of the embryo and in the maintenance of embryonic stem cell pluripotency. **Subunit:** Homodimer. Interacts with NR5A1, NR5A2, NR0B2 and with COPS2. **Subcellular Location:** Nucleus. Cytoplasm. Shuttles between the cytoplasm and nucleus. Homodimers exits in the cytoplasm and in the nucleus. **DISEASE:** Defects in NR0B1 are the cause of X-linked adrenal hypoplasia congenital (XL-AHC) [MIM:300200]; also known as X-linked Addison disease (AHX). XL-AHC is a developmental disorder of the adrenal gland that results in profound hormonal deficiencies and is lethal if untreated. It is characterized by the absence of the permanent zone of the adrenal cortex and by a structural disorganization of the glands. Hypogonadotropic hypogonadism (HHG) is frequently associated with this disorder. HHG is a condition resulting from or characterized by abnormally decreased gonadal function, with retardation of growth and sexual development. Defects in NR0B1 are the cause of 46,XY sex reversal type 2 (SRXY2) [MIM:300018]. It is a condition characterized by male-to-female sex reversal in the presence of a normal 46,XY karyotype. Note=XY individuals with a duplication of part of the short arm of the X chromosome and an intact SRY gene develop as females. The single X chromosome in these individuals does not undergo X-chromosome inactivation; therefore, these individuals presumably carry 2 active copies of genes, including the NR0B1 gene, in the duplicated region. Individuals with deletion of this region develop as males. Genes within the dosage-sensitive sex reversal region are, therefore, not essential for testis development, but, when present in a double dose, interfere with testis formation.

Similarity:
Belongs to the nuclear hormone receptor family. NR0 subfamily.

SWISS:
P51843

Gene ID:
190

Database links:

Entrez Gene: 450140 Chimpanzee

Entrez Gene: 190 Human

Entrez Gene: 11614 Mouse

Omim: 300473 Human

SwissProt: Q9BG97 Chimpanzee

SwissProt: P51843 Human

SwissProt: Q61066 Mouse

Unigene: 268490 Human

Unigene: 5180 Mouse

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