

# Rabbit Anti-CYP11A1/FITC Conjugated antibody

# SL3608R-FITC

Product Name:	Anti-CYP11A1/FITC
Chinese Name:	FITC标记 <b>的</b> 细 <b>胞色素</b> P450 11A1 <b>抗体</b>
Alias:	Cholesterol 20 22 desmolase; Cholesterol desmolase; Cholesterol monooxygenase (side chain cleaving); Cholesterol side chain cleavage enzyme; Cholesterol side chain cleavage enzyme mitochondrial; Cholesterol side-chain cleavage enzyme; CP11A HUMAN; CYP11A; CYP11A1; CYPXIA1; Cytochrome P450 11A1;
	Cytochrome P450 11A1 mitochondrial; Cytochrome P450 family 11 subfamily A polypeptide 1; Cytochrome P450 subfamily XIA; Cytochrome P450(scc); Cytochrome P450C11A1; mitochondrial; P450SCC; Steroid 20 22 lyase.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human, Mouse, Rat,
Applications:	IF=1:50-200 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	53/57kDa
Cellular localization:	The cell membrane Mitochondrion
Form:	Lyophilized or Liquid
Concentration:	lmg/ml
immunogen:	KLH conjugated synthetic peptide derived from human CYP11A1/P450SCC
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.
Product Detail:	background: This gene encodes a member of the cytochrome P450 superfamily of enzymes. The

cytochrome P450 proteins are monooxygenases which catalyze many reactions involved in drug metabolism and synthesis of cholesterol, steroids and other lipids. This protein localizes to the mitochondrial inner membrane and catalyzes the conversion of cholesterol to pregnenolone, the first and rate-limiting step in the synthesis of the steroid hormones. Two transcript variants encoding different isoforms have been found for this gene. The cellular location of the smaller isoform is unclear since it lacks the mitochondrial-targeting transit peptide. [provided by RefSeq, Jul 2008]

#### Function:

Catalyzes the side-chain cleavage reaction of cholesterol to pregnenolone.

## **Subcellular Location:**

Mitochondrion membrane.

#### **DISEASE:**

Defects in CYP11A1 are the cause of adrenal insufficiency congenital with 46,XY sex reversal (AICSR) [MIM:613743]. A rare disorder that can present as acute adrenal insufficiency in infancy or childhood. ACTH and plasma renin activity are elevated and adrenal steroids are inappropriately low or absent; the 46,XY patients have female external genitalia, sometimes with clitoromegaly. The phenotypic spectrum ranges from prematurity, complete underandrogenization, and severe early-onset adrenal failure to term birth with clitoromegaly and later-onset adrenal failure. Patients with congenital adrenal insufficiency do not manifest the massive adrenal enlargement typical of congenital lipoid adrenal hyperplasia.

## Similarity:

Belongs to the cytochrome P450 family.

# Database links:

Entrez Gene: 1583Human

Omim: 118485Human

SwissProt: P05108Human

Unigene: 303980Human

# **Important Note:**

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