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CYP11A1 Recombinant Rabbit mAb

Catalog # AP100076

Product Information

Application	WB, IHC-P, IHC-F, IF, IP, E
Primary Accession	P05108
Other Accession	P05108
Reactivity	Rat, Mouse
Predicted	Human
Host	Rabbit
Clonality	Recombinant
Calculated MW	60102
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CYP11A1
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 cytochrome P450 family.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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]

Additional Information

Gene ID	1583
Other Names	CYP11A; CYPXIA1; P450SCC; D9Ertd411e; Scc; cscc; P450(scc); CP11A_HUMAN; CYP11A1; Cholesterol desmolase; Cytochrome P450 11A1; Cytochrome P450(scc); 1.14.15.6; CP11A_MOUSE; CP11A_RAT; Cyp11a-1; cytochrome P450

	family 11 subfamily A member 1; cytochrome P450, subfamily XIA (cholesterol side chain cleavage); cytochrome P450, family 11, subfamily A, polypeptide 1; cholesterol monooxygenase (side-chain-cleaving)
Dilution	WB=1:1000-2000,IHC-P=1:100-500,IF=1:100-500,IP=1:50-100,ELISA=1:5000-10000,IHC-F=1:100-500
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	CYP11A1 {ECO:0000303|PubMed:21636783, ECO:0000312|HGNC:HGNC:2590}
Function	A cytochrome P450 monooxygenase that catalyzes the side-chain hydroxylation and cleavage of cholesterol to pregnenolone, the precursor of most steroid hormones (PubMed:21636783). Catalyzes three sequential oxidation reactions of cholesterol, namely the hydroxylation at C22 followed with the hydroxylation at C20 to yield 20R,22R- hydroxycholesterol that is further cleaved between C20 and C22 to yield the C21-steroid pregnenolone and 4-methylpentanal (PubMed:21636783). Mechanistically, uses molecular oxygen inserting one oxygen atom into a substrate and reducing the second into a water molecule. Two electrons are provided by NADPH via a two-protein mitochondrial transfer system comprising flavoprotein FDXR (adrenodoxin/ferredoxin reductase) and nonheme iron-sulfur protein FDX1 or FDX2 (adrenodoxin/ferredoxin) (PubMed:21636783).
Cellular Location	Mitochondrion inner membrane {ECO:0000250|UniProtKB:P14137}; Peripheral membrane protein. Note=Localizes to the matrix side of the mitochondrion inner membrane. {ECO:0000250|UniProtKB:P14137}

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