

CYP11A1 Rabbit pAb

CYP11A1 Rabbit pAb Catalog # AP52081

Product Information

Application WB
Primary Accession P05108
Reactivity Rat
Host Rabbit
Clonality Polyclonal
Calculated MW 60102
Physical State Liquid

Immunogen KLH conjugated synthetic peptide derived from human CYP11A1/P450SCC

Epitope Specificity 321-420/521

Isotype IgG

Purity affinity purified by Protein A

Buffer 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.

SUBCELLULAR LOCATION Mitochondrion membrane.

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 Cholesterol side-chain cleavage enzyme, mitochondrial, 1.14.15.6, CYPXIA1,

Cholesterol desmolase, Cytochrome P450 11A1, Cytochrome P450(scc),

CYP11A1 {ECO:0000303 | PubMed:21636783,

ECO:0000312 | HGNC:HGNC:2590}

Dilution WB=1:500-5000

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}

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]

References

Chung B.-C., et al. Proc. Natl. Acad. Sci. U.S.A. 83:8962-8966(1986).

Morohashi K., et al.J. Biochem. 101:879-887(1987).

Ota T., et al. Nat. Genet. 36:40-45(2004).

Zody M.C., et al. Nature 440:671-675(2006).

Mural R.J., et al. Submitted (SEP-2005) to the EMBL/GenBank/DDBJ databases.

Images

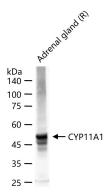


figure) probed with CYP11A1 polyclonal antibody, unconjugated (AP52081) at 1:1000 dilution and 4°C overnight incubation. Followed by conjugated secondary antibody incubation at r.t. for 60 min.

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