

CYP11B2 Rabbit pAb

Catalog # AP99584

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P19099
Other Accession	P19099
Host	Rabbit
Clonality	Polyclonal
Calculated MW	57560
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CYP11B2
Epitope Specificity	251-350/503
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 CYP11B2 are the cause of corticosterone methyloxidase type 1 deficiency (CMO-1 deficiency) [MIM:203400]; also known as aldosterone deficiency due to defect in 18-hydroxylase or aldosterone deficiency I. CMO-1 deficiency is an autosomal recessive disorder of aldosterone biosynthesis. There are two biochemically different forms of selective aldosterone deficiency be termed corticosterone methyloxidase (CMO) deficiency type 1 and type 2. In CMO-1 deficiency, aldosterone is undetectable in plasma, while its immediate precursor, 18-hydroxycorticosterone, is low or normal. Defects in CYP11B2 are the cause of corticosterone methyloxidase type 2 deficiency (CMO-2 deficiency) [MIM:610600]. CMO-2 is an autosomal recessive disorder of aldosterone biosynthesis. In CMO-2 deficiency, aldosterone can be low or normal, but at the expense of increased secretion of 18-hydroxycorticosterone. Consequently, patients have a greatly increased ratio of 18-hydroxycorticosterone to aldosterone and a low ratio of corticosterone to 18-hydroxycorticosterone in serum. Defects in CYP11B2 are a cause of familial hyperaldosteronism type 1 (FH1) [MIM:103900]. It is a disorder characterized by hypertension, variable hyperaldosteronism, and abnormal adrenal steroid production, including 18-oxocortisol and 18-hydroxycortisol. There is significant phenotypic heterogeneity, and some individuals never develop hypertension. Note=The molecular defect causing hyperaldosteronism familial type 1 is an anti-Lepore-type fusion of the CYP11B1 and CYP11B2 genes. The hybrid gene has the promoting part of CYP11B1, ACTH-sensitive, and the coding part of CYP11B2.
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. The enzyme has steroid 18-hydroxylase activity to synthesize aldosterone and 18-oxocortisol as well as steroid 11 beta-hydroxylase activity. Mutations in this gene cause corticosterone methyl oxidase deficiency.

Additional Information

Gene ID	1585
Other Names	ALDOS; CPN2; CYP11B; CYP11BL; CYPXIB2; P-450C18; P450C18; P450aldo; Cyp11b-2; Cp45as; Cyp11b3; RNCP45AS; C11B2_HUMAN; CYP11B2; Aldosterone synthase (ALDOS); Aldosterone-synthesizing enzyme; Corticosterone 18-monooxygenase, CYP11B2; Cytochrome P-450Aldo; Cytochrome P-450C18; Steroid 11-beta-hydroxylase, CYP11B2; Steroid 18-hydroxylase; C11B2_MOUSE; Cytochrome P450C11; C11B2_RAT; Cytochrome P450(11beta, aldo) (P450(11beta, aldo)); Cytochrome P450-Aldo-1; cytochrome P450 family 11 subfamily B member 2; cytochrome P450, subfamily XIB (steroid 11-beta-hydroxylase), polypeptide 2; cytochrome P450, family 11, subfamily B, polypeptide 2; steroid 11-beta-monooxygenase
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=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	CYP11B2 {ECO:0000303 PubMed:1346492, ECO:0000312 HGNC:HGNC:2592}
Function	A cytochrome P450 monooxygenase that catalyzes the biosynthesis of aldosterone, the main mineralocorticoid in the human body responsible for salt and water homeostasis, thus involved in blood pressure regulation, arterial hypertension, and the development of heart failure (PubMed:11856349, PubMed:12530636, PubMed:1518866, PubMed:15356073, PubMed:1594605, PubMed:1775135, PubMed:22446688, PubMed:23322723, PubMed:9814482, PubMed:9814506). Catalyzes three sequential oxidative reactions of 11-deoxycorticosterone (21-hydroxyprogesterone), namely 11-beta hydroxylation, followed by two successive oxidations at C18 yielding 18-hydroxy and then 18-oxo intermediates (that would not leave the enzyme active site during the consecutive hydroxylation reactions), ending with the formation of aldosterone (PubMed:11856349, PubMed:12530636, PubMed:1518866, PubMed:1594605, PubMed:1775135, PubMed:22446688, PubMed:23322723, PubMed:9814506). Can also produce 18-hydroxycortisol and 18-oxocortisol, derived from successive oxidations of cortisol at C18, normally found at very low levels, but significantly increased in primary aldosteronism, the most common form of secondary hypertension (PubMed:15356073, PubMed:9814482). 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:11856349, PubMed:1594605, PubMed:23322723, PubMed:23685396, PubMed:9814506). Besides its role in the biosynthesis of mineralocorticoids and glucocorticoids, it can also participate in the androgen metabolic pathway, converting testosterone (17beta-hydroxy-4-androsten-3-one) to 11beta,17beta-dihydroxyandrost-4-

ene-3-one (PubMed:[23685396](#)). Can also use progesterone as substrate (PubMed:[33189850](#)).

Cellular Location

Mitochondrion inner membrane {ECO:0000250 | UniProtKB:P14137};
Peripheral membrane protein {ECO:0000250 | UniProtKB:P14137}

Tissue Location

Expressed sporadically in the zona glomerulosa (zG) of the adrenal cortex (conventional zonation), as well as in aldosterone-producing cell clusters (APCCs) composed of morphological zG cells in contact with the capsule (variegated zonation)

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.