

# PCBD1 (N-12): sc-165196

## BACKGROUND

PCBD1 (pterin-4  $\alpha$ -carbinolamine dehydratase), also known as PCD, PHS, DCOH (dimerization cofactor of hepatocyte nuclear factor 1  $\alpha$ ) or PCBD, is a component of the phenylalanine hydroxylase (PAH) system and participates in tetrahydrobiopterin biosynthesis. More specifically, PCBD1 catalyzes the dehydration of pterin-4 $\alpha$ -carbinolamine (4-OH-BH4) to quinonoid dihydrobiopterin (q-BH2), an essential reaction for the regeneration of 6(R)-L-erythro-5,6,7,8-tetrahydrobiopterin (6(R)BH4). In addition, PCBD1 can homodimerize and, in this dimer, can function as a transcriptional activator cofactor for HNF-1 $\alpha$ . Mutations in the gene encoding PCBD1 lead to an accumulation of 4-OH-BH4 which subsequently produces 7-BH4 (a potent inhibitor of PAH), and may result in primapterinuria. Patients with primapterinuria (a mild form of phenylketonuria (PKU) exhibit both hyperphenylalaninemia (HPA) and excretion of 7-substituted pterins.

## REFERENCES

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## CHROMOSOMAL LOCATION

Genetic locus: PCBD1 (human) mapping to 10q22.1; Pcbd1 (mouse) mapping to 10 B4.

## SOURCE

PCBD1 (N-12) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of PCBD1 of human origin.

## PRODUCT

Each vial contains 200  $\mu$ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-165196 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

PCBD1 (N-12) is recommended for detection of PCBD1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000); non cross-reactive with PCBD2.

PCBD1 (N-12) is also recommended for detection of PCBD1 in additional species, including canine.

Suitable for use as control antibody for PCBD1 siRNA (h): sc-90559, PCBD1 siRNA (m): sc-152050, PCBD1 shRNA Plasmid (h): sc-90559-SH, PCBD1 shRNA Plasmid (m): sc-152050-SH, PCBD1 shRNA (h) Lentiviral Particles: sc-90559-V and PCBD1 shRNA (m) Lentiviral Particles: sc-152050-V.

Molecular Weight of PCBD1: 12 kDa.

Positive Controls: C32 whole cell lysate: sc-2205.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## STORAGE

Store at 4° C, \*\*DO NOT FREEZE\*\*. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

## RESEARCH USE

For research use only, not for use in diagnostic procedures.

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.