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SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

mail@szabo-scandic.com

www.szabo-scandic.com

[linkedin.com/company/szaboscandic](https://www.linkedin.com/company/szaboscandic) 

PCBD1 (E-7): sc-518184

BACKGROUND

PCBD1 (pterin-4 α -carbinolamine dehydratase), also known as PCD, PHS, DCOH (dimerization cofactor of hepatocyte nuclear factor 1 α) 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 α -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 α . 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

1. Thöny, B., et al. 1998. Hyperphenylalaninemia with high levels of 7-biopterin is associated with mutations in the PCBD gene encoding the bifunctional protein pterin-4 α -carbinolamine dehydratase and transcriptional coactivator (DCoH). *Am. J. Hum. Genet.* 62: 1302-1311.
2. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 126090. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
3. Schallreuter, K.U., et al. 2003. Molecular evidence that halo in Sutton's naevus is not vitiligo. *Arch. Dermatol. Res.* 295: 223-228.
4. Schallreuter, K.U., et al. 2004. Activation/deactivation of acetylcholinesterase by H₂O₂: more evidence for oxidative stress in vitiligo. *Biochem. Biophys. Res. Commun.* 315: 502-508.

CHROMOSOMAL LOCATION

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

SOURCE

PCBD1 (E-7) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 25-49 of PCBD1 of human origin.

PRODUCT

Each vial contains 200 μ g IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

PCBD1 (E-7) is available conjugated to agarose (sc-518184 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-518184 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-518184 PE), fluorescein (sc-518184 FITC), Alexa Fluor® 488 (sc-518184 AF488), Alexa Fluor® 546 (sc-518184 AF546), Alexa Fluor® 594 (sc-518184 AF594) or Alexa Fluor® 647 (sc-518184 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-518184 AF680) or Alexa Fluor® 790 (sc-518184 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

PCBD1 (E-7) is recommended for detection of PCBD1 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], 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).

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.

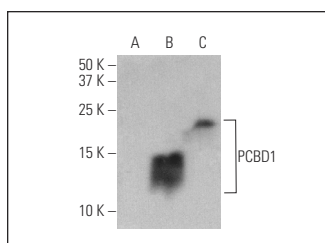
Molecular Weight of PCBD1: 12 kDa.

Positive Controls: PCBD1 (m): 293T Lysate: sc-122420 or human PCBD1 transfected 293T whole cell lysate.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



PCBD1 (E-7): sc-518184. Western blot analysis of PCBD1 expression in non-transfected 293T: sc-117752 (A), mouse PCBD1 transfected 293T: sc-122420 (B) and human PCBD1 transfected 293T (C) whole cell lysates. Detection reagent used: m-IgG₁ BP-HRP: sc-525408.

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 for detailed protocols and support products.