

PDC-E2 (C-9): sc-271352

BACKGROUND

Primary biliary cirrhosis (PBC) is a chronic, destructive autoimmune liver disease characterized by the presence of antimitochondrial autoantibodies in patient's serum and T cell-mediated destruction of the biliary epithelial cells lining the small intrahepatic bile ducts. Patient sera are characterized by a high frequency (greater than 95%) of autoantibodies directed to a mitochondrial antigen, identified as the E2 component of the pyruvate dehydrogenase multienzyme complex (PDC-E2). PDC-E2 contains both an amino-terminal lipoyl-bearing domain and a carboxy-terminal catalytic domain. The human sequence preserves the Glu-Thr-Asp-Lys-Ala motif of the lipoyl-bearing site. Two conformationally alternative forms of the PDC-E2 protein have been revealed by immunoblotting. The immunodominant autoepitopes of the autoantigens correspond to the inner lipoyl domain. A significant number of asymptomatic patients found to have antibodies to PDC-E2 are at high risk of developing primary biliary cirrhosis.

REFERENCES

1. Coppel, R.L., et al. 1988. Primary structure of the human M2 mitochondrial autoantigen of primary biliary cirrhosis: dihydrolipoamide acetyltransferase. *Proc. Natl. Acad. Sci. USA* 85: 7317-7321.
2. Thekkumkara, T.J., et al. 1988. Nucleotide sequence of a cDNA for the dihydrolipoamide acetyltransferase component of human pyruvate dehydrogenase complex. *FEBS Lett.* 240: 45-48.

CHROMOSOMAL LOCATION

Genetic locus: DLAT (human) mapping to 11q23.1; Dlat (mouse) mapping to 9 A5.3.

SOURCE

PDC-E2 (C-9) is a mouse monoclonal antibody raised against amino acids 231-390 mapping within an internal region of PDC-E2 of human origin.

PRODUCT

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

APPLICATIONS

PDC-E2 (C-9) is recommended for detection of PDC-E2 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg 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 PDC-E2 siRNA (h): sc-40813, PDC-E2 siRNA (m): sc-40814, PDC-E2 shRNA Plasmid (h): sc-40813-SH, PDC-E2 shRNA Plasmid (m): sc-40814-SH, PDC-E2 shRNA (h) Lentiviral Particles: sc-40813-V and PDC-E2 shRNA (m) Lentiviral Particles: sc-40814-V.

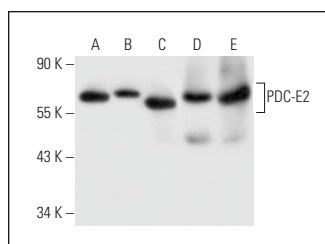
Molecular Weight of PDC-E2: 70 kDa.

Positive Controls: KNRK whole cell lysate: sc-2214, mouse liver extract: sc-2256 or Caki-1 cell lysate: sc-2224.

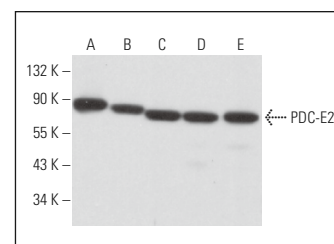
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ 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-IgGκ BP-FITC: sc-516140 or m-IgGκ 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



PDC-E2 (C-9): sc-271352. Western blot analysis of PDC-E2 expression in KNRK (A) and Caki-1 (B) whole cell lysates and mouse liver (C), rat kidney (D) and mouse heart (E) tissue extracts.



PDC-E2 (C-9): sc-271352. Western blot analysis of PDC-E2 expression in SK-BR-3 (A), NIH/3T3 (B), c4 (C) and C2C12 (D) whole cell lysates and rat liver tissue extract (E).

SELECT PRODUCT CITATIONS

1. Lebigoit, E., et al. 2017. Impact of mutations within the [Fe-S] cluster or the lipoyl acid biosynthesis pathways on mitochondrial protein expression profiles in fibroblasts from patients. *Mol. Genet. Metab.* 122: 85-94.
2. Kwak, C.H., et al. 2019. Huzhangoside A suppresses tumor growth through inhibition of pyruvate dehydrogenase kinase activity. *Cancers* 11: 712.
3. Kwak, C.H., et al. 2020. Ilimaquinone induces the apoptotic cell death of cancer cells by reducing pyruvate dehydrogenase kinase 1 activity. *Int. J. Mol. Sci.* 21: 6021.

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.