

PDC-E2 (A-9): sc-365276

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

- 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.
- 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.
- Klein, R., et al. 1993. Sera from patients with tuberculosis recognize the M2a-epitope (E2 subunit of pyruvate dehydrogenase) specific for primary biliary cirrhosis. *Clin. Exp. Immunol.* 92: 308-316.
- Chen, Q.Y., et al. 1993. Antibody to two forms of dihydrolipoamide acetyltransferase (PDC-E2) in primary biliary cirrhosis. *Liver* 13: 130-135.
- Howard, M.J., et al. 1998. Three-dimensional structure of the major autoantigen in primary biliary cirrhosis. *Gastroenterology* 115: 139-146.

CHROMOSOMAL LOCATION

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

SOURCE

PDC-E2 (A-9) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 83-117 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.

Blocking peptide available for competition studies, sc-365276 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

RESEARCH USE

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

APPLICATIONS

PDC-E2 (A-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), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

PDC-E2 (A-9) is also recommended for detection of PDC-E2 in additional species, including canine, porcine and avian.

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.

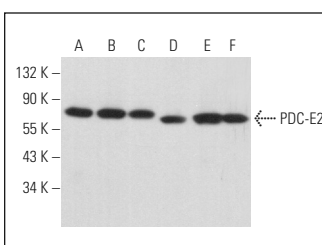
Molecular Weight of PDC-E2: 70 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, MCF7 whole cell lysate: sc-2206 or Sol8 cell lysate: sc-2249.

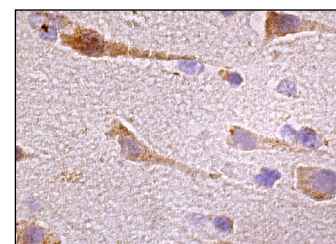
RECOMMENDED SUPPORT REAGENTS

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. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



PDC-E2 (A-9): sc-365276. Western blot analysis of PDC-E2 expression in HeLa (A), MCF7 (B), SJRH30 (C), Sol8 (D) and L6 (E) whole cell lysates and rat postnatal heart tissue extract (F).



PDC-E2 (A-9): sc-365276. Immunoperoxidase staining of formalin fixed, paraffin-embedded human brain tissue showing cytoplasmic staining of neuronal cells.

STORAGE

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