



SZABO SCANDIC

Part of Europa Biosite

Produktinformation



Forschungsprodukte & Biochemikalien



Zellkultur & Verbrauchsmaterial



Diagnostik & molekulare Diagnostik



Laborgeräte & Service

Weitere Information auf den folgenden Seiten!
See the following pages for more information!



Lieferung & Zahlungsart

siehe unsere [Liefer- und Versandbedingungen](#)

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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) 



PDC-E2 (m): 293T Lysate: sc-122447

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., McNeilage, L.J., Surh, C.D., Van de Water, J., Spithill, T.W., Whittingham, S. and Gershwin, M.E. 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., Ho, L., Wexler, I.D., Pons, G., Liu, T.C. and Patel, M.S. 1988. Nucleotide sequence of a cDNA for the dihydrolipoamide acetyltransferase component of human pyruvate dehydrogenase complex. *FEBS Lett.* 240: 45-48.
3. Klein, R., Wiebel, M., Engelhart, S. and Berg, P.A. 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.
4. Chen, Q.Y., Rowley, M.J. and Mackay, I.R. 1993. Antibody to two forms of dihydrolipoamide acetyltransferase (PDC-E2) in primary biliary cirrhosis. *Liver* 13: 130-135.
5. Howard, M.J., Fuller, C., Broadhurst, R.W., Perham, R.N., Tang, J.G., Quinn, J., Diamond, A.G. and Yeaman, S.J. 1998. Three-dimensional structure of the major autoantigen in primary biliary cirrhosis. *Gastroenterology* 115: 139-146.
6. Palmer, J.M., Diamond, A.G., Yeaman, S.J., Bassendine, M.F. and Jones, D.E. 1999. T cell responses to the putative dominant autoepitope in primary biliary cirrhosis (PBC). *Clin. Exp. Immunol.* 116: 133-139.
7. Quaranta, S., Van de Water, J., Ishibashi, H., Rosina, F., Coppel, R., Uibo, R. and Gershwin, M.E. 1999. The immunopathogenesis of primary biliary cirrhosis. *Hepatogastroenterology* 46: 3041-3047.
8. Kisand, K.E., Metskula, K., Kisand, K.V., Kivik, T., Gershwin, M.E. and Uibo, R. 2001. The follow-up of asymptomatic persons with antibodies to pyruvate dehydrogenase in adult population samples. *J. Gastroenterol.* 36: 48-54.

CHROMOSOMAL LOCATION

Genetic locus: Dlat (mouse) mapping to 9 A5.3.

PRODUCT

PDC-E2 (m): 293T Lysate represents a lysate of mouse PDC-E2 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

PDC-E2 (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive PDC-E2 antibodies. Recommended use: 10-20 µl per lane.

Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. 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.