

Properdin (h): 293T Lysate: sc-159880

BACKGROUND

The serum complement system (SCS), containing over 30 glycoproteins, influences physiological mechanisms of the body in response to immune complex (the classical pathway), carbohydrate (the lectin pathway) or bacterial (alternative pathway) initiation. Properdin, also known as complement factor P (CFP), PFC, BFP or PFD, is a secreted glycoprotein that participates in positively regulating the alternative pathway of the SCS. Properdin exists as a cyclic polymer with six Thrombospondin type 1 domains and binds to C3 and C5 convertase complexes (C3bBb and (C3b)_nBb) functioning to assist in their stabilization. Properdin is also required for the deposition of C3b onto the surface of pathogens. Mutations in the gene encoding Properdin can result in Properdin deficiency (PFD), a disease characterized by higher susceptibility to bacterial infections.

REFERENCES

1. Fredrikson, G.N., Gullstrand, B., Westberg, J., Sjöholm, A.G., Uhlen, M. and Truedsson, L. 1998. Expression of Properdin in complete and incomplete deficiency: normal *in vitro* synthesis by monocytes in two cases with Properdin deficiency type II due to distinct mutations. *J. Clin. Immunol.* 18: 272-282.
2. Vuagnat, B.B., Mach, J. and Le Doussal, J.M. 2000. Activation of the alternative pathway of human complement by autologous cells expressing transmembrane recombinant Properdin. *Mol. Immunol.* 37: 467-478.
3. van den Bogaard, R., Fijen, C.A., Schipper, M.G., de Galan, L., Kuijper, E.J. and Mannens, M.M. 2000. Molecular characterisation of 10 Dutch Properdin type I deficient families: mutation analysis and X-inactivation studies. *Eur. J. Hum. Genet.* 8: 513-518.
4. Hartmann, S. and Hofsteenge, J. 2000. Properdin, the positive regulator of complement, is highly C-mannosylated. *J. Biol. Chem.* 275: 28569-28574.
5. Jelezarova, E., Vogt, A. and Lutz, H.U. 2000. Interaction of C3b₂-IgG complexes with complement proteins Properdin, Factor B and Factor H: implications for amplification. *Biochem. J.* 349: 217-223.
6. Perdikoulis, M.V., Kishore, U. and Reid, K.B. 2001. Expression and characterisation of the Thrombospondin type I repeats of human Properdin. *Biochim. Biophys. Acta* 1548: 265-277.
7. Bongrazio, M., Pries, A.R. and Zakrzewicz, A. 2003. The endothelium as physiological source of Properdin: role of wall shear stress. *Mol. Immunol.* 39: 669-675.
8. Hourcade, D.E. 2006. The role of Properdin in the assembly of the alternative pathway C3 convertases of complement. *J. Biol. Chem.* 281: 2128-2132.
9. Sprong, T., Roos, D., Weemaes, C., Neeleman, C., Geesing, C.L., Mollnes, T.E. and van Deuren, M. 2006. Deficient alternative complement pathway activation due to factor D deficiency by 2 novel mutations in the complement factor D gene in a family with meningococcal infections. *Blood* 107: 4865-4870.

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.

CHROMOSOMAL LOCATION

Genetic locus: CFP (human) mapping to Xp11.23.

PRODUCT

Properdin (h): 293T Lysate represents a lysate of human Properdin transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

Properdin (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive Properdin antibodies. Recommended use: 10-20 µl per lane.

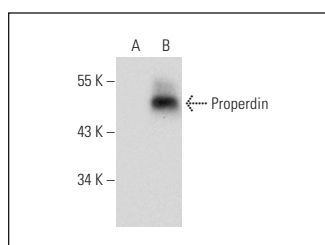
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

Properdin (M-184): sc-68367 is recommended as a positive control antibody for Western Blot analysis of enhanced human Properdin expression in Properdin transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

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.

DATA



Properdin (C-4): sc-365664. Western blot analysis of Properdin expression in non-transfected: sc-117752 (A) and human Properdin transfected: sc-159880 (B) 293T whole cell lysates.

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.