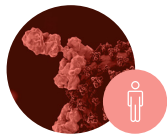


Product datasheet



ATP02

Proteinase 3 (cANCA) Native Antigen

Product name	Proteinase 3 (cANCA) antigen
Description	Purified native protein
Source material	Human neutrophils
Purity	>90% pure (by SDS-PAGE)

Antigen overview

Autoantibodies to neutrophil cytoplasmic antigens (ANCA) were first described in 1982 by Davies *et al.*¹ Autoantibodies staining the nuclei or the perinuclear zone of neutrophils by indirect immunofluorescence are referred to as pANCA whereas those giving a clear cytoplasmic fluorescence are referred to as cANCA².

The antigen recognised by most cANCA sera has been identified as proteinase 3 (also known as myeloblastin, p29 or AGP7)³⁻⁵. Autoantibodies to proteinase 3 are found in the sera of patients with active Wegener's granulomatosis⁶ and less often in other types of systemic vasculitis including microscopic polyangiitis, idiopathic crescentic glomerulonephritis, Churg-Strauss syndrome and polyarteritis nodosa⁷.

Proteinase 3 is a serine proteinase with proteolytic activity towards a range of substrates⁸. It is physiologically inhibited by α 1-antitrypsin⁹. The protein has been described as having antibiotic¹⁰ and growth-promoting¹¹ properties and causing

empyema when administered by tracheal insufflation¹². Proteinase 3 cDNA has been cloned and sequenced¹⁰, revealing the protein to be a basic 25 kDa glycoprotein of 228 amino acids¹³.

The protein reveals a triplet of bands in the range 29-32 kDa when subjected to denaturing SDS-electrophoresis, probably caused by differing degrees of glycosylation¹⁴. Although proteinase 3 shows significant homology to elastase and cathepsin G, cANCA sera do not cross react with these serine proteases¹⁵.

Autoantibodies to proteinase 3 appear to recognise conformational epitopes¹⁶ and have been reported to interfere with inactivation of the enzyme by α 1-antitrypsin¹⁷ and to also inhibit proteolytic activity itself¹⁷.

The use of purified proteinase 3 for the detection of cANCA by solid-phase ELISA has been described by several authors^{2,18-25}.

Storage conditions & handling

- Store at -65°C or below
- Avoid repeated freezing and thawing
- Mix before use and keep on ice

Ordering information

ATP02-02 - Proteinase 3 (cANCA) antigen (native) - 0.20 mg

ATP02-10 - Proteinase 3 (cANCA) antigen (native) - 1.0 mg

Custom vial sizes available on request

Applications

Reactive in Western Blot and ELISA with anti-PR3 antibodies or anti-PR3 positive patient serum

Product profile

Fig 1 AROTEC Proteinase 3 (cANCA) Antigen Titration

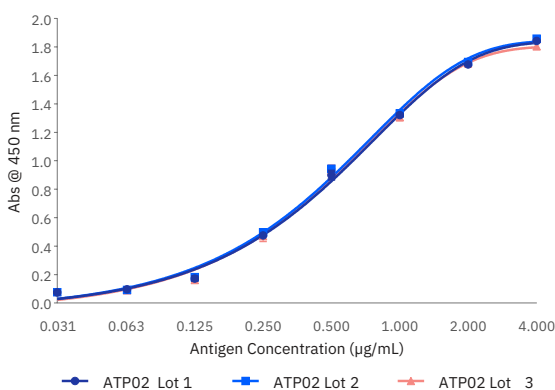


Fig 2

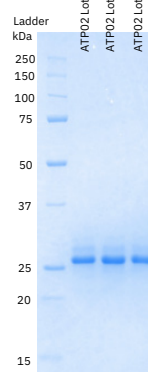


Fig 3

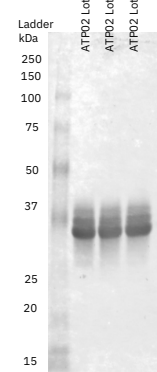


Fig.1. ELISA titration assay of ATP02 Proteinase 3 (cANCA) antigen using positive patient serum (1:100 dilution)

Fig.2. SDS-PAGE of ATP02 Proteinase 3 (cANCA) antigen; All lanes loaded with 1 µg protein concentration

Fig.3. Western Blot of ATP02 Proteinase 3 (cANCA) antigen with positive patient serum (1:300 dilution)

References

- Davies, D.J. *et al.* (1982) Br. Med. J. 285, 606
- Wiik, A. *et al.* (1994) Man. Biol. Markers of Dis. (Kluwer Acad.Publ.) A9:1-14
- Ludemann, J. *et al.* (1990) J. Exp. Med. 171, 357
- Jenne, D.E. *et al.* (1990) Nature 346, 520
- Gupta, S.K. (1990) Blood, 76, 2162
- Wiik, A. (1995) Semin. Clin. Immunol. 9, 5
- Kallenberg, C.G.M. *et al.* (1994) Kidney Int. 46, 1
- Kam, C.M. *et al.* (1992) FEBS Lett. 297, 119
- Dolman, K.M. *et al.* (1992) FEBS Lett. 314, 117
- Campanelli, D. *et al.* (1990) J. Exp. Med. 172, 1709
- Bories, D. *et al.* (1989) Cell 59, 959
- Kao, R.C. *et al.* (1988) J. Clin. Invest. 82, 1963
- Wieslander, J. & Wiik, A. (1994) Man. Biol. Markers of Dis.(Kluwer Acad. Publ.) B7:1-1-9
- Goldschmeding, R. *et al.* (1989) J. Clin. Invest. 84, 1577
- Nassberger, L. *et al.* (1993) Autoimmunity 14, 259
- Bini, P. *et al.* (1992) J. Immunol. 149, 1409
- Van den Wiel, A. *et al.* (1992) Clin. Exp. Immunol. 90, 409
- Ludemann, J. *et al.* (1988) J. Immunol. Meth. 114, 167
- Daha, M.R. *et al.* (1990) Neth. J. Med. 36, 117
- Kallenberg, C.G.M. & Rasmussen, N. (1990) Neth. J. Med. 36, 132
- Gaskin, G. *et al.* (1995) J. Immunol. Meth. 180, 25
- Stumann, L. & Wiik, A. (1997) J. Immunol. Meth. 206, 35
- Heegaard, N.H.H. *et al.* (1997) Anal. Biochem. 253, 259
- Hagen, E.C. *et al.* (1993) J. Immunol. Meth. 159, 1
- Hagen, E.C. *et al.* (1996) J. Immunol. Meth. 196, 1