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AM26220PU-N Monoclonal Antibody to PR3 / C-ANCA - Purified

Alternate names: AGP7, Leukocyte proteinase 3, MBN, Myeloblastin, NP-4, Neutrophil proteinase 4, P29, PR-3, PRTN3, Proteinase 3, Wegener autoantigen

Quantity: 0.1 mg

Concentration: 0.1 mg/ml

Background: PR3 is a major antigen recognized by autoantibodies directed against cytoplasmic proteins of neutrophilic granulocytes and monocytes (called anti-neutrophil cytoplasmic autoantibodies (ANCA)). ANCA are able to activate primed neutrophils to produce oxygen radicals and release lytic enzymes, including PR3. Proteinase 3 (PR3) was identified as the target antigen of ANCA in Wegener's granulomatosis (WG). ANCA directed against PR3 (PR3-ANCA) can interfere with the binding of PR3 to its physiological inhibitor alpha1-antitrypsin (alpha1-AT) and with the proteolytic activity of PR3. At the site of inflammation, PR3 can cleave the PR3-ANCA complex between these inhibiting ANCA and PR3 itself, leaving active PR3. Autoantibodies to PR3 are potent activators of the 5-lipoxygenase pathway in primed human neutrophils. Extracellular free arachidonic acid, as present at an inflammatory focus, synergizes with such autoantibodies to evoke full-blown lipid mediator generation, granule secretion and respiratory burst. Proteinase 3 (PR3) is a neutral serine proteinase, which is localized in the azurophilic granules of neutrophils and in granules of monocytes and can be detected in the membrane of secretory vesicles. PR3 degrades a number of extracellular matrix proteins such as elastin and inactivates human C1 inhibitor. Membrane-associated PR3 is also able to activate caspase-3 without triggering apoptosis of neutrophils, which is possibly a neutrophil survival mechanism. In addition, PR3 is involved in myeloid differentiation and is, therefore, also called myeloblastin.

Uniprot ID: P24158

NCBI: 9606

GeneID: 5657

Host / Isotype: Mouse / IgG1

Recommended Isotype Controls: SM10P (for use in human samples), AM03095PU-N

Clone: PR3-G2

Immunogen: A crude granule extract

Format:
State: Liquid 0.2 µm filtered Ig fraction
Purification: Protein G
Buffer System: PBS
Preservatives: 0.02% sodium azide
Stabilizers: 0.1% bovine serum albumin

Applications:
Western blot: The typical starting working dilution is 1:50.
Immunoassay.
Flow cytometry: The typical starting working dilution is 1:50.
Immunohistochemistry on frozen sections: The typical starting working dilution is 1:50.
Other applications not tested. Optimal dilutions are dependent on conditions and should be determined by the user.

Specificity:
Monoclonal antibody PR3G-2 reacts with human proteinase 3 (PR3), a 30 kDa protein.

Species Reactivity:
Tested: Human

Storage:
Store at 2 - 8 °C.
Shelf life: one year from despatch.

General Readings:
1. Van Der Geld YM, Limburg PC, Kallenberg CG. Characterization of monoclonal antibodies to proteinase 3 (PR3) as candidate tools for epitope mapping of human anti-PR3 autoantibodies. Clin Exp Immunol. 1999 Dec;118(3):487-96. PubMed PMID: 10594572.
2. Pederzoli M, Kantari C, Gausson V, Moriceau S, Witko-Sarsat V. Proteinase-3 induces procaspase-3 activation in the absence of apoptosis: potential role of this compartmentalized activation of membrane-associated procaspase-3 in neutrophils. J Immunol. 2005 May 15;174(10):6381-90. PubMed PMID: 15879139.