

AM32114PU-N**Monoclonal Antibody to PR3 / C-ANCA - Liquid****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

Clone:

PR3G-2

Immunogen:

Immunization of mice with a crude granule extract.

Format:**State:** Liquid purified Ig fraction (0.2 µm filtered)**Buffer System:** PBS**Preservatives:** 0,02% sodium azide**Stabilizers:** 0,1% BSA**Applications:****Flow Cytometry.****Immunohistochemistry on frozen sections.****Western Blot.****Immunoassay.**

It is recommended that users test the reagent and determine their own optimal dilutions.

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:

This antibody reacts to Proteinase 3 (PR3)

Species Reactivity:

Tested: Human.

Storage:

Store the antibody undiluted at 2-8°C for one month or (in aliquots) at -20°C for longer.

Avoid repeated freezing and thawing.

Shelf life: one year from despatch.