**AM26221PU-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 (so-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 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:** NP_002768.3  
**GeneID:** 5657  
**Host / Isotype:** Mouse / IgG1  
**Recommended Isotype Controls:** SM10P (for use in human samples), AM03095PU-N  
**Clone:** WGM2  
**Format:** Liquid 0.2 µm filtered Ig fraction  
**Purification:** Protein G Chromatography  
**Buffer System:** PBS  
**Preservatives:** None  
**Stabilizers:** 0.1% BSA  
**Applications:** Flow Cytometry, Immunoassays, Western blot.  
The typical starting working dilution is 1/10.
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**Functional assays:** Blocks the PR3 activity and partially inhibits the binding of human PR3-ANCA to PR3.

**Immunohistochemistry on Frozen and Paraffin Sections.**
Other applications not tested. Optimal dilutions are dependent on conditions and should be determined by the user.

**Specificity:**
This Monoclonal antibody WGM2 reacts with Human Proteinase 3 (PR3), a 30 kDa protein.

**Species Reactivity:**
**Tested:** Human

**Storage:**
Store undiluted at 2-8°C.
**DO NOT FREEZE!**
Shelf life: one year from despatch.

**General Readings:**