Autoantibodies against Bactericidal/Permeability-Increasing Protein (BPI)

Bactericidal/permeability-increasing protein (BPI) is an endogenous antibiotic protein thas was originally identified as a constituent of the azurophilic granules in neutrophilic granulocytes/polymorphonuclear leukocytes (PMNs). BPI is released into the phagosome to directly destroy Gram-negative bacteria by increasing their membrane permeability. In addition, BPI has been found to be released into the extracellular space and to bind to the lipid A region of lipopolysaccharides (LPS) of Gram-negative bacteria. This binding neutralizes LPS, which is an endotoxin and a strong inducer of the inflammatory response, and has been shown to promote opsonization and bacterial phagocytosis. Two functional domains have been identified in BPI. While the cationic N-terminal domain exerts the anti-bacterial activity and LPS binding, both the N- and C-terminal domain are required for the opsonic function.

Importantly, BPI has also been found to be expressed in mucosal epithelial cells underlining its function in the prevention of respiratory infections.

Anti-neutrophil cytoplasmic antibodies (ANCA) against the C-terminus of BPI have been recognized in vasculitis, cystic fibrosis (CF), TAP (transporter associated with antigen presentation) deficiency, inflammatory bowel disease, autoimmune hepatitis, sclerosing cholangitis and - comparatively rare - in systemic vasculitides. BPI autoantibodies represent a subgroup of ANCA that give mostly rise to a cytoplasmic pattern (cANCA) in indirect immunofluorescence assays using ethanol fixed granulocytes.

A common clinical feature of the above mentioned diseases is the chronic or profuse exposure of the patient to Gram-negative bacteria, and in turn the necessity to neutralize and dispose bacterial endotoxins like LPS, which involves BPI. In CF, BPI autoantibodies have been reported to be associated with *Pseudomonas aeruginosa* colonizing the lung and lung damage. A similar association with BPI autoantibodies may also exist in case of TAP deficiency. At least in CF it can be speculated that BPI autoantibodies represent a marker of chronic endobronchial infection and sustained inflammatory response.

To complement the offered ANCA antigens myeloperoxidase (MPO) and proteinase 3 (PR3), DIARECT's product portfolio contains native human bactericidal/permeability-increasing protein (BPI) purified from polymorphonuclear leukocytes of peripheral human blood.

Ordering Information		
18500 18501	Myeloperoxidase (MPO; non recombinant)	0.1 mg 1.0 mg
18600 18601	Proteinase 3 (PR3; non recombinant)	0.1 mg 1.0 mg
19200 19201	BPI (non recombinant)	0.1 mg 1.0 mg

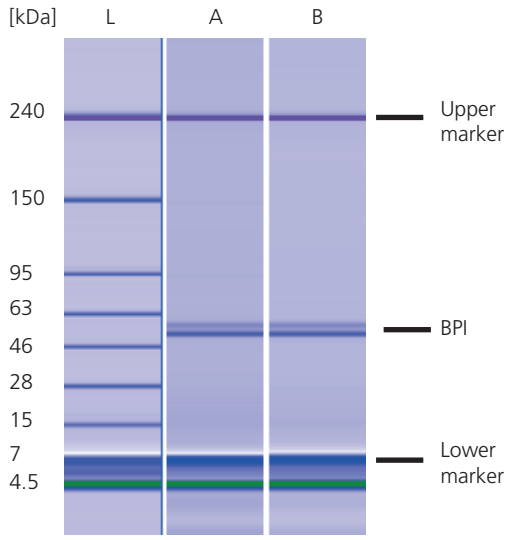

Figure: Electrophoretic analyses of two independent lots of native, non-recombinant BPI isolated from human neutrophils (A and B). The loading buffer added to the BPI samples contained an upper and lower marker. The molecular weight of the protein standards included in the size ladder (L) are indicated on the left.

References:

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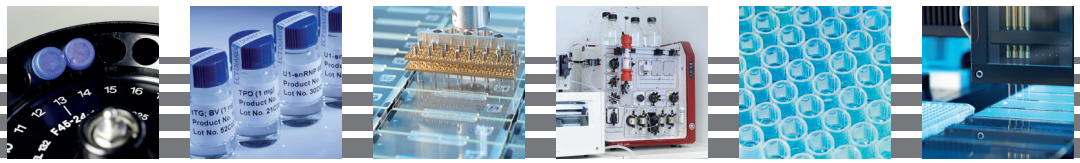

DIARECT AG · Bötzinger Str. 29 B · 79111 Freiburg · Germany
Tel. +49 761 47979-0 · Fax +49 761 47979-29 · orders@diarect.com · www.diarect.com