

C3 Proactivator, for *BN™ Series* and *Atellica® NEPH 630* nephelometers

General information: structure, function ...

The complement system Factor B (FB), also known as C3 Proactivator (C3PA), Properdin Factor B (PFB) or Glycine-rich Beta Glycoprotein (GBG), is a glycoprotein with an approximate molecular weight of 93 KDa, consisting of a single peptide chain, which is one of the proteins in the complement cascade alternative pathway (Complement Factors).

Factor B binds to plasma C3 which, in small amounts, is activated by spontaneous hydrolysis (C3i). Factor D (FD) acts on the complex (C3iB) and breaks down Factor B (FB) into Ba and Bb, forming a C3 convertase of the alternative pathway (C3iBb) which cleaves more C3 into C3a and C3b, and which in turn can bind to more FB thus forming C3 convertase (C3bBb). This spontaneous activation of the alternative pathway remains in a dormant state since fluid phase C3b is rapidly inactivated.

While body cells are protected by the action of Factor H and other regulatory proteins, in the presence of invasive microorganisms (such as many gram negative and some gram positive bacteria, certain viruses, yeasts and parasites) part of spontaneously generated C3b binds to cell membranes. This C3b then stabilizes and binds to FB, which by action of FD generates C3 convertase (C3bBb), which is in turn stabilized by the action of Properdin (Factor P), thus creating a feedback loop that activates the alternative pathway. More C3b binds to convertase C3bBb, forming C5 convertase in the alternative pathway (C3bBb3b), which continues with the common part of complement activation.

Clinical Significance

Although Factor B deficiency is rare, when it does occur, it compromises the activation of the complement alternative pathway which is essential in defence against bacterial infections (especially *Neisseria*).

Reduced levels of Factor B normally indicate activation of the complement alternative pathway. Measurement of FB is helpful when diagnosing certain kinds of kidney diseases (such as chronic glomerulonephritis or lupus nephritis), skin diseases (such as dermatitis herpetiformis or pemphigus vulgaris), rheumatoid arthritis, sickle cell anaemia, as well as certain other infections, such as gram-negative infections.

Assay Performances and Characteristics

- ➔ **Nephelometric Immunoassay (NIA)**, for their use on *BN™ Series and Atellica® NEPH 630* nephelometers from *Siemens Healthcare* (*BN™* and *Atellica®* are a registered trademark of Siemens Healthcare).
- ➔ Standardized to the *WHO's International Ref. Preparation for human serum complement Factors* (code: W1032).
- ➔ Reagents, Calibrator and Controls ready to use in barcode-identified containers.

Catalogue

N C3Pa Kit

REF TD-42720

▽ 100 test

EAN/GTIN: 8434477212086

SMN: 10873732

Contains Reagents, Calibrator and Controls (2 levels)

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Assay also available for other analytical platforms. For further information, please contact the Customer Support Service at support@3diag.com