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32-3372: C1q Native Protein

Alternative Name : Component C1q,Complement C1q,Complement Component C1q,C1q.
Immunogen Information : Human Plasma.

Description

Source : Human Plasma. Human Complement Component C1q produced in Human plasma having a molecular mass of 410 kDa. C1q is the first component of the classical pathway of complement activation. C1q along with the enzymatically active components C1r and C1s forms the C1 complex. When C1 binds to immunoglobulins in the form of immune complexes, it leads to activation of C1r and C1s proteases and a further activates the classical pathway of complement. C1q is a glycoprotein that belongs to the collectin family, having a molecular weight of about 410-462 kDa. C1q is a hexamer composed of globular heads attached to collagen-like triple-helix tails. The globular heads of C1q exclusively bind to the CH2 domain of IgG molecules or the CH3 domain of IgM. Each heavy chain of the immunoglobulin molecule contains a single binding site for C1q. Given that C1q must bind to no less than two heavy chains in order to alter its conformation and activate C1r and C1s, its activation follows only after binding to immunoglobulins in the form of immune complexes bound to multivalent antigens. C1q's main physiological role is in the clearance of immune complexes and apoptotic bodies from the organism. Interruption of this process may lead to development of autoimmunity. Individuals with genetic deficiencies of C1q or other components of the classical pathway are at risk to develop SLE. C1q specifically binds to apoptotic bodies of human keratinocytes, vascular endothelial cells and lymphocytes. Complement components C1q and bound C3 mediate the clearance of apoptotic bodies. Hence, C1q may advance the clearance of autoantigens, avoiding stimulation of the immune system. Nonetheless, an extended exposition of the immune system to the neoepitope exposed on C1q molecules bound to immune complexes or apoptotic bodies could ultimately lead to an autoimmune response against C1q itself and to an altered complement function. C1q deficiency may also lead to disruption of the negative selection of autoreactive B cells. C1q along with other specific recognition proteins bind to the highly conserved lupus antigens (dsDNA and nuclear proteins) and activate the complement system. Autoantibodies against C1q (anti-C1q) are found in a number of autoimmune and infectious diseases like glomerulonephritis (GN) and lupus erythematosus (SLE), these antibodies are significant in clinical practice due to their negative predictive value.

Product Info

Amount :	200ug / 1 mg
Purification :	Greater than 95.0% as determined by SDS-PAGE.
Content :	In sterile filtered 10 mM HEPES and 300mM NaCl, pH 7.2. Concentration is lot dependent.
Storage condition :	Human C1q is stable at 4°C if entire vial will be used within 2-4 weeks. Store, frozen below -20°C for longer periods of time. For long term storage it is recommended to add a carrier protein (0.1% HSA or BSA). Avoid multiple freeze-thaw cycles.

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