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C1QC Mouse mAb

Catalog # AP100290

Product Information

Application	WB, IHC-P, IF, IP, E
Primary Accession	P02747
Other Accession	P02747
Predicted	Human
Host	Mouse
Clonality	Monoclonal
Calculated MW	25774
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human C1QC
Epitope Specificity	101-155/245
Isotype	IgG1, k
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 1 C1q domain. Contains 1 collagen-like domain.
SUBUNIT	C1 is a calcium-dependent trimolecular complex of C1q, R and S in the molar ration of 1:2:2. C1q subcomponent is composed of nine subunits, six of which are disulfide-linked dimers of the A and B chains, and three of which are disulfide-linked dimers of the C chain.
Post-translational modifications	O-linked glycans consist of Glc-Gal disaccharides bound to the oxygen atom of post-translationally added hydroxyl groups.
DISEASE	Defects in C1QC are a cause of complement component C1q deficiency (C1QD) [MIM:613652]. A rare defect resulting in C1 deficiency and impaired activation of the complement classical pathway. C1 deficiency generally leads to severe immune complex disease with features of systemic lupus erythematosus and glomerulonephritis.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	C1q, a subcomponent of the classical complement pathway, is composed of nine subunits that mediate classical complement activation and thereby play an important role in the immune response. Six of these subunits are disulfide-linked dimers of chains A and B, while three of these subunits, designated C1q-A through C1q-C, are disulfide-linked dimers of chain C. The presence of receptors for C1q on effector cells modulates its activity, which may be antibody-dependent or independent. Macrophages are the primary source of C1q, while anti-inflammatory drugs as well as cytokines differentially regulate expression of the mRNA, as well as the protein. However, its ability to modulate the interaction of platelets with collagen and immune complexes suggests C1q influences homeostasis as well as other immune activities, and perhaps thrombotic complications resulting from immune injury. Defects in C1q-A, C1q-B and C1q-C cause inactivation of the classical pathway, leading to a rare genetic disorder characterized by lupus-like symptoms.

Additional Information

Gene ID	714
Other Names	C1Q-C; C1QD3; C1QG; Adib; Ciqc; C1QC_HUMAN; C1QC; C1QC_MOUSE; C1QC_RAT; complement C1q C chain; complement component 1, q subcomponent, gamma polypeptide; complement component 1, q subcomponent, C chain
Dilution	WB=1:100-1000,IP=1:20-100,IHC-P=1:50-500,ELISA=1:30-3000,IF=1:50-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	C1QC {ECO:0000303|PubMed:1706597, ECO:0000312|HGNC:HGNC:1245}
Function	Core component of the complement C1 complex, a multiprotein complex that initiates the classical pathway of the complement system, a cascade of proteins that leads to phagocytosis and breakdown of pathogens and signaling that strengthens the adaptive immune system (PubMed:12847249, PubMed:19006321, PubMed:24626930, PubMed:29449492, PubMed:3258649, PubMed:34155115, PubMed:6249812, PubMed:6776418). The classical complement pathway is initiated by the C1Q subcomplex of the C1 complex, which specifically binds IgG or IgM immunoglobulins complexed with antigens, forming antigen-antibody complexes on the surface of pathogens: C1QA, together with C1QB and C1QC, specifically recognizes and binds the Fc regions of IgG or IgM via its C1q domain (PubMed:12847249, PubMed:19006321, PubMed:24626930, PubMed:29449492, PubMed:3258649, PubMed:6776418). Immunoglobulin-binding activates the proenzyme C1R, which cleaves C1S, initiating the proteolytic cascade of the complement system (PubMed:29449492). The C1Q subcomplex is activated by a hexamer of IgG complexed with antigens, while it is activated by a pentameric IgM (PubMed:19706439, PubMed:24626930, PubMed:29449492). The C1Q subcomplex also recognizes and binds phosphatidylserine exposed on the surface of cells undergoing programmed cell death, possibly promoting activation of the complement system (PubMed:18250442).
Cellular Location	Secreted. Cell surface. Note=Specifically binds IgG or IgM immunoglobulins complexed with antigens, forming antigen-antibody complexes on the surface of pathogens.

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