

CERTIFICATE OF ANALYSIS – TECHNICAL DATA SHEET

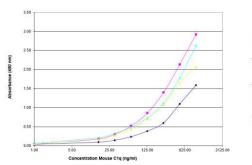
Product name C1q, Mouse, clone JL-1

| Catalog number | HM1096-5MG | | |
|----------------|-------------------------------|---------------|------------|
| Lot number | - | Expiry date | - |
| Volume | - | Amount | 5 mg |
| Formulation | $0.2 \ \mu m$ filtered in PBS | Concentration | >0.5 mg/ml |
| Host Species | Mouse IgG2b | Conjugate | None |
| Endotoxin | <24 EU/mg | Purification | Protein G |
| Storage | 4°C | | |

Application notes

| | IHC-F | IHC-P | IF | FC | FS | IA | IP | w |
|-------------|-------|-------|----|----|----|-------|----|---|
| Reference # | 1 | | 1 | | 1 | 1,2,4 | | 3 |
| Yes | ٠ | | • | | • | ٠ | | • |
| No | | | | | | | | |
| N.D. | | • | | • | | | • | |

N.D.= Not Determined; IHC = Immuno histochemistry; F = Frozen sections; P = Paraffin sections; IF = Immuno Fluorescence; FC = Flow Cytometry; FS = Functional Studies; IA = Immuno Assays; IP = Immuno Precipitation; W = Western blot



| | Amount C1q | Ong | 10ng | Song |
|----------------------|--------------------------|-----|------|------|
| Human EDTA plasma | PBS | | - | * |
| | Denaturing sample buffer | | _ | |
| CD1 mouse serum | PBS | | • | • |
| | Denaturing sample buffer | | | |
| Human EDTA plasma | PBS | | • | • |
| | Denaturing sample buffer | | | |
| CD1 mouse serum | PBS | | • | |
| | Denaturing sample buffer | | | |

 $\ensuremath{\mathsf{IA}}\xspace$: HM1096 was used as a detection antibody in different concentrations.

Dot blot with HM1096 with mouse serum and human plasma with different amounts of C1q.

Dilutions to be used depend on detection system applied. It is recommended that users test the reagent and determine their own optimal dilutions. The typical starting working dilution is 1:50.

- W: Mouse bone-marrow derived macrophages; non-reduced; ~125 kDa (Ref 1); reduction with 2-mercaptoethanol destroys BM8 antigen.
- IHCF: Antibody JL-1 was used to stain tissue sections which were fixed in acetone. As positive control a polyclonal anti-C1q antibody
 was used and as negative control an isotype matched monoclonal antibody (Ref.1).
- FS: Antibody JL-1 was administered to mice resulting in depletion of circulating C1q, glomerular deposition of C1q and induction of anti-C1q autoantibodies in susceptible mice. As a negative control an isotype matched monoclonal antibody was used (Ref.1).
- Positive control: Spleen and kidney tissue of wild-type mice (Ref.1); Negative control: Spleen and kidney tissue of C1q -/- mice (Ref.1).

| General Information | | |
|---------------------|--|--|
| Description | The monoclonal antibody JL-1 recognizes the collagen-like region (CLR) of mouse C1q, a 459 kDa molecule consisting of three individual polypeptide chains. The antibody has been generated by immunization of C1q-/- C57BL/6 mice with purified mouse C1g. | |
| | C1q forms together with C1r and C1s the C1 macromolecule, the first component of the classical complement pathway. Interaction of immune complexes with C1q induces a conformational change within the C1 complex, which results in activation of the classical pathway. C1q functions as recognition unit by binding to the heavy chain of IgG or IgM (Fc gamma and Fc micro) provided that the immunoglobulins are bound to their antigen. Furthermore, C1q can also recognize molecular patterns associated with pathogens and it can bind to apoptotic blebs, where it activates the | |
| | classical complement pathway and mediates phagocytosis. As such, C1q promotes the clearance of apoptotic cells and subsequent exposure of auto antigens, thereby preventing stimulation of the immune system. | |
| | C1q is predominantly produced by macrophages but also by follicular dendritic cells, interdigitating cells and cells of the monocyte-macrophage lineage. C1q deficiency has a profound effect on host defence and clearance of immune | |

| | complexes. Absence of C1q may cause autoimmunity by impairment of the clearance of apoptotic cells. Inherited C1q deficiency is also associated with the development of systemic lupus erythematosus (SLE). The monoclonal antibody JL-1 is reactive with the collagen-like region (CLR) only, which is the same region to which autoantibodies in mice and humans are binding. Anti-C1q autoantibodies deposit in glomeruli together with C1q but induce overt renal disease only in the context of glomerular immune complex disease. This provides an explanation why anti-C1q antibodies are especially pathogenic in patients with SLE. | | | |
|-------------------|---|--|--|--|
| Immunogen | Purified mouse C1q | | | |
| Aliases | Complement component C1q, Complement C1q subcomponent subunit A | | | |
| Cross reactivity | Human: Yes; Rat: Yes. | | | |
| References | Trouw, L et al; Anti-C1q autoantibodies deposit in glomeruli but are only pathogenic in combination with glomerular C1q-containing immune complexes. J Clin Invest 2004, <i>114</i>: 679 Li, M et al; Development of a humanized C1q A chain knock-in mouse: assessment of antibody independent beta- amyloid induced complement activation. Mol Immunol 2008, <i>45</i>: 3244 Erlich, P et al; Complement protein C1q forms a complex with cytotoxic prion protein oligomers. J Biol Chem 2010, <i>285</i>: 19267 Kiriakidis, S et al; Complement C1q is hydroxylated by collagen prolyl 4 hydroxylase and is sensitive to off-target inhibition by prolyl hydroxylase domain inhibitors that stabilize hypoxia-inducible factor. Kid Int 2017, <i>92</i>: 900. | | | |
| Storage&stability | Product should be stored at 4°C. Under recommended storage conditions, product is stable for at least one year. | | | |
| Precautions | For research use only. Not for use in or on humans or animals or for diagnostics. It is the responsibility of the user to comply with all local/state and federal rules in the use of this product. Hycult Biotech is not responsible for any patent infringements that might result from the use or derivation of this product. | | | |

We hereby certify that the above-stated information is correct and that this product has been successfully tested by the Quality Control Department. This product was released for sale according to the existing specifications. This document has been produced electronically and is valid without a signature.

Approved by Manager of QC Brenda Teunissen

Date 07/10/2019

Do you have any questions or comments regarding this product? Please contact us via support@hycultbiotech.com.