Complement factors

General:

Essential functions of the complement system are their participation in the defense of infectious agents and in preventing autoimmune diseases. To our current knowledge the complement system consists of approx. 20 glycoproteins in inactive forms and receptors on blood cells. As with the coagulation system, the complement system is activated stepwise (cascade): the inactive protein is transferred into the activated one. The classical way of activation starts by binding C1q to antibodies, which must have reacted a priori with a specific antigen on the erythrocyte surface.

The alternative way starts with activation of C3 without activating C1, C2 and C4. Antibody binding shows no effect on the alternative way, but e.g. bacterial endotoxines or cobra poison do. The result of the complete activation is a lysis complex, which leads to cytolysis, bacteriolysis, and hemolysis or to virus neutralization. Common causes of complement increase are systemic infections, non-infectious chronic inflammation conditions as well as physiological conditions (e.g. pregnancy). The hypocomplementemia is an important indicator for the diagnosis of immune-complex disorders.

The following tests are available:

- **C1q Complement binding**

General:

C1q is a 400kDa protein formed from 18 peptide chains in 3 subunits of 6. Each 6-peptide subunit consists of a Y-shaped pair of triple peptide helices joined at the stem and ending in a globular non-helical head. The C1q complex is potentially multivalent for attachment to the complement fixation sites of immunoglobulin. The sites are on the CH2 domain of IgG and probably on the CH4 domain of IgM. The appropriate peptide sequence of the complement fixing site might become exposed following complexing of the immunoglobulin, or the sites might always be available, but might require multiple attachment by C1q with critical geometry in order to achieve the necessary avidity.

Clinical significance are deficiencies of the subunit of the first complement factors C1q, C1r and/or C1s. The most important defect is the C1qdeficiency, which often is associated with Systemic Lupus Erythematodes (SLE). Decreased levels are also detected with hypogammaglobulinemia, hypocomplementary urticaria-vasculitis and severe immune deficiency (SCID).

Indication	Diseases with complement activation, including the classic way of the complement activation, susp. of C1q deficiency, urticaria vasculitis with complement deficiency
Material:	1 ml serum
TAT:	8-15 days*
Method:	NEPH
Units:	mg/dl
Ref.- range:	13.0 – 32.0

- **C2 Complement**

General:

Diseases with classical complement activation, suspicion for C2 deficiency, immuncomplex associated diseases, hereditary angioneurotic oedema

Indication:	Diseases with classical complement activation
Material:	1 ml serum
TAT:	10-14 days*
Method:	RID
Units:	%
Ref.- range:	80 – 120

- **C3 Complement**

Indication:	Suspicion of complement activation, includes the classical as well as the alternative pathway of complement activation
Material:	1 ml serum
Stability:	8 days at 2 to 8°C
TAT:	same day, FML
Method:	TURB
Units:	mg/dl
Ref.- range:	see report

- **C3 Complement in Aspirate**

Material: 1 ml aspirate
TAT: 7-10 days*
Method: NEPH
Units: mg/dl
Ref.- range: 90-180

- **C3 Complement in seminal fluid**

Material: 1 ml seminal fluid
TAT: 7-10 days*
Method: TURB
Units: mg/dl
Ref.- range: see report

- **C3 Nephritis factor**

General:

In membrane proliferative glomerulonephritis autoantibodies can occur against C3-convertase of the alternative pathway of the complement activation, the so-called C3-nephritis factor. Blocking of C3-convertase leads to unrestrained transformation of C3, sometimes up to complete consumption of the protein. The resulting decreased humoral immunity increases the infection risk. Effects on kidney transplants must be considered. Simultaneous decrease of the hemolytic total complement activity with severely reduced C3 points to the presence of C3-nephritis factor.

Material: 1 ml serum
TAT: 7-10 days*
Method: IEP
Units: mg/dl
Ref.- range: see report

- **C3 Proactivator**

Material: 1 ml serum, Frozen
TAT: 7-10 days*
Method: RID
Units: mg/dl
Ref.- range: 17.0 - 42.0

- **C3d Complement**

Indication: Diseases with complement activation, including the classical as well as alternative pathway of the complement activation
Material: 1 ml serum, Frozen
TAT: 7-10 days*
Method: EIA
Units: U/ml
Ref.- range: 6.3-10.9
Comments: Decreased in glomerulonephritis, liver cell damage, SLE, polyarthritis rheumatica; increased in bacterial infections and non-specific inflammatory processes.

- **C4 Complement**

Material: 1 ml serum, Frozen
TAT: same day, FML
Stability: 8 days at 2 to 8°C
Method: TURB
Units: mg/dl
Ref.- range: 12.0 - 36.0
Comments: Decreased in angioneurotic edema, hereditary C4 deficiency, glomerulonephritis, SLE, alpha-1-antitrpysin deficiency, vasculitis, liver cell damage, polyarthritis rheumatica

- **C4 Complement in aspirate**

Material: 1 ml aspirate
TAT: 7-10 days*
Method: NEPH
Units: mg/dl
Ref.- range: see report

- **C5 Complement**

Material: 1 ml serum
TAT: 10-14 days*
Method: RID
Units: %
Ref.- range: 80 - 120

- **C6 Complement**

Material: 1 ml serum
TAT: 10-14 days*
Method: RID
Units: %
Ref.- range: 80 - 120

- **C7 Complement**

Material: 1 ml serum
TAT: 10-14 days*
Method: RID
Units: %
Ref.- range: 80 - 120

- **C8 Complement**

Material: 1 ml serum
TAT: 10-14 days*
Method: RID
Units: %
Ref.- range: 80 - 120

- **C9 Complement**

Material: 1 ml serum
TAT: 10-14 days*
Method: RID
Units: %
Ref.- range: 80 - 120

- **Total Hemolytic Complement (CH50)**

General :

Screening test of the total activity of the complement system. If the hemolytic activity is missing or if it is strongly reduced, a complement deficiency has to be considered. This could also occur as a consequence of substantial activation. Increased local activation of the complement system can lead to tissue damage or to anaphylactic reactions.

Indication: Detection of activation of the complement system, monitoring patients at risk (e.g. after polytraumas, burning, sepsis and transplantations);
Material: 1 ml serum, frozen
Preanalytics: After blood collection, allow the blood to coagulate completelely at room temperature (Do not cool!). Decant the serum immediately after centrifugation, freeze the serum at minus 20°C (-20°C) and store/transport the sample frozen.
TAT: 7-10 days*
Method: TURB
Units: U/ml
Ref.- range: 41.7 - 95.1

For complete list of laboratory test offered at Freiburg Medical Laboratory, please visit http://www.fml-dubai.com/parameter-listings/