Complement Deficiency Testing

**INDICATIONS FOR TESTING**

Suspected complement deficiency (recurrent bacterial infections, especially respiratory infections or infection with *Neisseria* species; autoimmune disorders, especially systemic lupus erythematosus)

**ORDER**

- CH50 assay for total hemolytic complement
- AH50 assay for alternative pathway hemolytic activity
- Also consider evaluation for immunoglobulin disorders

**NORMAL CH50 and AH50**

High suspicion for complement deficiency

**ORDER**

- Mannose Binding Lectin (MBL)

**LOW OR ABSENT CH50**

Normal AH50

Classical pathway component deficiency or disorder of complement consumption

Consider complement component concentration or functional testing for C1, C2, and/or C4

**LOW OR ABSENT MBL**

Lectin pathway component deficiency

**LOW OR ABSENT AH50**

Normal CH50

Alternative pathway component deficiency

Consider any or all of the following:
- Properdin concentration or functional testing
- Factor B, D concentration testing

**SINGLE COMPONENT LOW**

Suggests hereditary deficiency; consider genetic testing

**MULTIPLE COMPONENTS LOW**

Suggests disorder associated with complement consumption

**TERMINAL PATHWAY COMPONENT DEFICIENCY OR DISORDER OF COMPLEMENT CONSUMPTION**

Consider any or all of the following:
- C3, C5, C6, C7, C8, C9 concentration or functional testing
- Factor H, I concentration testing

**CLASSICAL PATHWAY COMPONENT DEFICIENCY OR DISORDER OF COMPLEMENT CONSUMPTION**

Consider any or all of the following:
- C1, C2, C4 concentration or functional testing

**FOR PATIENTS TAKING C5 INHIBITOR DRUGS (EG, ECUILIZUMAB, RAVULIZUMAB), CONSIDER CH50, AH50, C5 CONCENTRATION, AND C5 FUNCTIONAL TESTING TO MONITOR COMPLEMENT BLOCKAGE AND ASSESS EFFICACY OF TREATMENT WITH C5 INHIBITORS.**

Refer to C5 Inhibitors Drug Monitoring Panel on the ARUP Laboratory Test Directory.

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