Deficiency of complement components

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Primary immunodeficiency

- Disorders of all individual components of complement,
- rare disorders,
- C1, C2, C3 and C4 disorders manifest as SLE-like immunocomplex diseases; combined with purulent infections,
- C1 inhibitor defects clinically severe → **hereditary angioedema**: uncontrolled complement activation occurs after various stimuli → local edema; airway swelling can be fatal,
- disorders in receptors for complement components include **LAD I syndrome** (affects CR3),
- in children with recurrent bacterial and fungal infections, **a deficiency of mannose-binding protein (lectin)** (MBL), which initiates the lectin pathway of complement activation, has been identified; in adults it is not manifested (compensated by other mechanisms).

Secondary immunodeficiency

- Reduction of complement components during its depletion, in case of synthesis failure,
- complement consumption in immunocomplex diseases, septic conditions,
- in severe liver disease,
  - normalization of liver functions → normalization of complement,
- transient consumption of C4 in attacks of hereditary angioedema.

Sources

- HOŘEJŠÍ, Václav – BARTŮŇKOVÁ, Jiřina. *Základy imunologie.* 3. edition. Praha : Triton, 2008. 280 pp. ISBN 80-7254-686-4.